Acral Fibromyxoma Treated with Reconstruction

M. Margoulis · D. Yeremenko · R. Wollstein

Received: 14 December 2012 / Accepted: 8 February 2013 / Published online: 17 February 2013
© Society of the Hand & Microsurgeons of India 2013

Keywords Acral Fibromyxoma · Benign tumour · Digital · Fracture

Introduction

Acral fibromyxoma is a benign soft tissue tumor that has been recently defined pathologically as a separate entity [1, 2]. Initially described by Fetsch et al. in 2001, it is characterized clinically by a slow-growing solitary tumor with a predilection for the subungual or periungual region of the hands and feet [1, 3]. In a large recent series, the diagnosis was not suggested by the clinician in the vast majority of patients, (122/124 cases) [1]. Due to the absence of long-term follow up information and the primary nature of the data regarding this entity, we suggest a conservative surgical approach when treating these patients.

Case Report

A 72-year-old female presented with a history of a mass in the distal phalanx of her ring finger that had been growing slowly over many years. She described periodical pain in her finger over the years but sought no treatment. There was no history of trauma or previous surgery in the finger.

Physical examination revealed a subcutaneous mass in the radial aspect of distal phalanx with destruction of the nail above the mass. There were no signs of acute inflammation or significant tenderness. She had slightly limited range of motion in the distal interphalangeal joint (DIP) most likely due to osteoarthritis that was present in all of the finger DIP joints. Radiographs revealed scalloping of the radial cortex of the distal phalanx with a sclerotic border. (Fig. 1) An ultrasound examination was performed demonstrating a 1 cm² solid mass.

The mass was removed under local anesthesia. No exsanguination was performed. The tumor involved the radial aspect of the distal phalanx both dorsally and volarly in the pulp. It seemed to include the radial side of the DIP joint as well. During removal, the phalanx that was already very thin fractured completely. A decision was made to avoid a disarticulation pending the pathology results. A small K-wire was inserted to maintain alignment of the fragments. The postoperative course was uneventful. The wire was removed at 6 weeks though union was not clear on radiographs.

Pathology Multiple fragments of white fibro-fatty tissue, about 2.5 cm² in volume, consistent with a superficial acral fibromyxoma. The tumor cells stained positive for CD34 and negative for S100, SMA and EMA. Assessment of margins could not be made. (Fig. 2)

Discussion

Acral Fibromyxoma, though recently characterized in a large collected series of 124 patients, remains a relatively unknown entity for the hand surgeon, despite the fact that the majority of cases occur in the distal fingertips [1]. This case was characteristic in presentation occurring as a solitary slow growing mass in the tip of a finger near the nail apparatus (97 % of the tumors described by Hollmann

M. Margoulis · R. Wollstein
Department of Orthopedic Surgery, Division of Hand and Upper
Extremity Surgery, Carmel Medical Center, Haifa, Israel

D. Yeremenko
Department of Pathology, Carmel Medical Center, Haifa, Israel

R. Wollstein (✉)
Department of Orthopedic Surgery and the Department of Plastic
Surgery, University of Pittsburgh, 3550 Terrace St,
Pittsburgh, PA 15261, USA
e-mail: ronitwollstein@gmail.com

Fig. 1 Posteroanterior (PA) and Lateral radiograph of the finger. The scalloping is clearly visualized. There is radiographic evidence of osteoarthrosis in the distal interphalangeal joint

Fig. 2 Hematoxylin and Eosin (H&E) stained slide x200 magnification. The tumor is composed of bland, spindle-shaped or stellate cells in a myxoid and collagenous stroma with scattered small vascular channels

et al.) [1]. In their series, pain was variable. Our patient described episodic pain throughout a long period of time.

During surgery, the tumor demonstrated an infiltrative pattern which is in tandem with 80 % of the Hollman series [1]. In their series, bone involvement was documented in 36 % of the cases with radiographic descriptions of an erosion or lytic lesion. Our case seems to differ from the typical pattern in the significant amount of bony involvement. A fracture necessitating fixation infers significant bone loss. Perhaps this was due to the long history of the tumor or from over aggressive curettage.

Since the tumor involved the bone and both the dorsal and volar aspect of the distal phalanx as well as the joint, a disarticulation was a reasonable surgical option. In the series described by Fetsch et al., 4/37 fingers underwent partial or complete amputation [3]. At this point the literature does not provide clear guidelines as to the type of excision that should be performed. There is a clear possibility of local recurrence when not completely excised and no incidence of remote metastases was described, but it is not clear if and how aggressively to address bone and joint involvement as well as soft tissue infiltration [1–3]. Due to the lack of long-term evidence, we suggest that a more conservative approach is justified at this time though because of the possibility of local recurrence follow-up should be continued.

Conflicts of interest The authors have no conflicts to disclose

References

1. Hollmann TJ, Bovee JV, Fletcher CD (2012) Digital fibromyxoma (superficial acral fibromyxoma): a detailed characterization of 124 cases. *Am J Surg Pathol* 36(6):789–798
2. Fanti PA, Dika E, Piraccini BM, Infusino SD, Baraldi C, Misciali C (2011) Superficial acral fibromyxoma: a clinicopathological and immunohistochemical analysis of 12 cases of a distinctive soft tissue tumor with a predilection for the fingers and toes. *G Ital Dermatol Venereol* 146(4):283–287
3. Fetsch JF, Laskin WB, Miettinen M (2001) Superficial acral fibromyxoma: a clinicopathologic and immunohistochemical analysis of 37 cases of a distinctive soft tissue tumor with a predilection for the fingers and toes. *Hum Pathol* 32(7):704–714