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Clinical case

## Desmoid tumor of the forearm. Reconstructive surgery and functional result

*Tumeur desmoïde de l'avant-bras. Résultats fonctionnels après excision–reconstruction en un temps*

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### Abstract

Functional results after resection–reconstruction operated at the same time. Desmoid tumors, also known as aggressive fibromatosis, are benign locally aggressive tumors with a high rate of recurrence. Most authors recommend surgical treatment with wide-free margin. Achieving margins of normal tissue around an upper extremity lesion without creating significant functional compromise is frequently difficult. Therefore, functional reconstructive surgery is important, considering that for most patients treated for these tumors, the life expectancy is high and considering also that wide resection can affect the function and the aesthetics as well. We present a case of aggressive fibromatosis in the proximal third of the forearm treated by wide resection and reconstructive surgery in one single procedure, with an acceptable functional result with no evidence of recurrence at 3 years of follow-up.

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*Keywords:* Desmoid tumor; Reconstructive surgery; Aggressive fibromatosis

### Résumé

Résultats fonctionnels après excision–reconstruction en un temps. Les tumeurs desmoïdes, ou fibromatose agressive, sont bénignes mais localement agressives avec un taux important de récurrence. La plupart des auteurs recommandent un traitement chirurgical avec résection large extramarginale. Nous présentons un cas de tumeur desmoïde au niveau du tiers proximal de l'avant-bras traité par résection étendue et chirurgie reconstructrice dans un même temps, avec un bon résultat fonctionnel et sans évidence de récurrence à trois ans. Compte tenu de l'espérance de vie, d'une part, et du préjudice esthétique et fonctionnel de l'excision, d'autre part, ce cas illustre l'intérêt d'une reconstruction fonctionnelle en un temps.

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*Mots clés :* Tumeur desmoïde ; Chirurgie reconstructrice ; Fibromatose agressive

### 1. Introduction

Desmoid tumors, also known as aggressive fibromatosis, are benign locally aggressive tumors with a high rate of recurrence. The patients often present with a deeply sited firm mass with or without pain. Most authors recommend surgical treatment with wide-free margins while many others also use additional adjuvant therapies, usually radiation [1–3].

Achieving margins of normal tissue around an upper extremity lesion without creating significant functional compromise is often difficult. Therefore, functional recon-

structive surgery is important considering that for most patients treated for these tumors, the life expectancy is high and considering also that wide resection can affect the function as well as the aesthetics.

We report the case of a desmoid tumor in the proximal third of a forearm treated by wide resection and reconstructive surgery in the same procedure.

### 2. Case report

A 23-year-old woman had a history of congenital dislocation of the radial head in her right elbow; she underwent surgical resection at another institution. Four years later, she was reoperated because of the presence of a tumor in the surgical

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area; the diagnosis found a desmoid tumor. One year after this procedure, she attended our hospital complaining of pain in the right elbow and wrist, limited flexion and extension in the elbow, posterior interosseous nerve paresis and a mass in the proximal third of the forearm where the two previous surgeries were done. During physical examination, several limitations were found in the range of motion. The flexion–extension of the elbow was 125–90 degrees, the pronation–supination was 45–0 degrees, the flexion of the wrist was 30 degrees and the extension was 10 degrees.

Plain radiographs showed heterotopic ossification between the anterior aspect of the radius and the distal humerus. Magnetic resonance imaging (MRI) was performed, with a 1.5-T Magnetom Vision unit (Siemens, Erlangen, Germany). This assessment showed a soft-tissue mass involving the posterior compartment of the forearm with low signal in T1-weighted and higher signal in T2-weighted (Fig. 1). A biopsy was done, reporting fibromatosis as a result.

A wide surgical resection of the tumor was done (Fig. 2). The mass compromised the belly muscle of the anconeus, extensor *pollicis brevis*, abductor *pollicis longus*, extensor *digitorum communis*, extensor *indicis*, extensor *digiti minimi* and extensor *carpi ulnaris*; all of them relevant of the posterior compartment. During surgery, it was found that the *brachioradialis* muscle had been previously resected, and extensor *carpi radialis brevis* had to be removed because it was also infiltrated by the tumor. As the posterior interosseous nerve was involved in the tumor, it had to be removed as well as the heterotopic ossification. Anterior arthrolysis was performed to improve elbow extension.

Tendon transfers were done as reconstructive surgery. Flexor *carpi radialis* was transferred to the extensor *digitorum communis*, extensor *indicis* and extensor *digiti minimi*. *Palmaris longus* was transferred to the extensor *pollicis brevis* and the abductor *pollicis longus*. Tenodesis was performed between the extensor *carpi ulnaris* remanent and the extensor

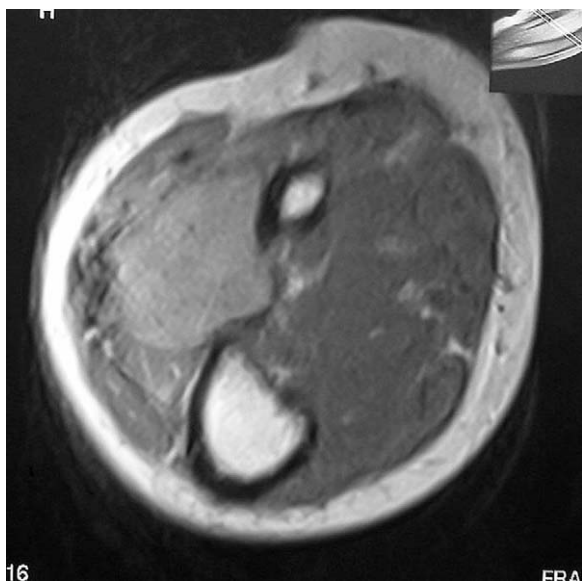


Fig. 1. The MRI showed a soft-tissue mass involving the posterior compartment of the forearm.

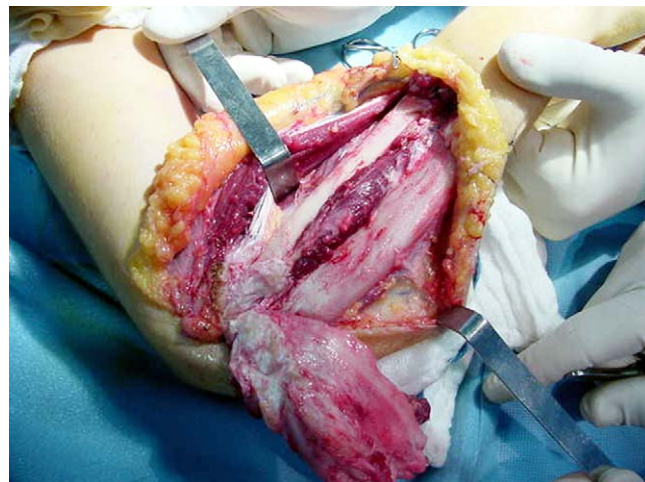


Fig. 2. The resection of the tumor included the posterior compartment of the forearm with the interosseous nerve.

*carpi radialis longus*, to prevent radial deviation of the hand. Due to the articular incongruence at the distal radioulnar joint, the Sauvé-Kapandji procedure was done to improve pronosupination and ease the pain.

Radiation therapy was administered as postsurgical treatment in dose of 50 Gy in 2 Gy daily fractions.

The pathologic anatomy showed tumor-free margins.

At 3 years of follow-up, the patient has 130–60 degrees of flexion–extension of the elbow, 45–0 degrees of pronation–supination and the flexion–extension of the wrist is 25–50 (Fig. 3 A, B). The grip strength measured with the Jamar dynamometer (Model 0030J4, Clifton, NJ) was 22 kg (75% of the contralateral side). The functional evaluation was performed using the revised 30-point functional classification system established by the International Society of Limb Salvage and the Musculoskeletal Tumor Society [4], and the Disabilities of the Arm, Shoulder and Hand (DASH) score [5]. The result of the first score was 3 and the second 19 points.

The patient returned to her usual job and at the time MRI showed no signs of recurrence of the tumor.

### 3. Discussion

We present the case of an aggressive fibromatosis of the forearm treated with wide resection and reconstructive surgery operated in one single procedure with an acceptable functional result with no evidence of recurrence at 3 years of follow-up.

The natural history of desmoid tumor remains an enigma [6,7]. Merchant et al. [8], analyzed 189 patients with primary extremity and trunk desmoid tumors treated and followed within a single institution in order to determine which factors influence the disease-free survival. None of the prognostic factors analyzed, including age, gender, depth of tumor, size of tumor or tumor site, were significant for predicting local recurrence.

Although surgical “trauma” has been implied as an etiologic factor of the desmoid tumors, there is still uncertainty about its significance [1,2]. Enzinger and Shiraki [1] reported

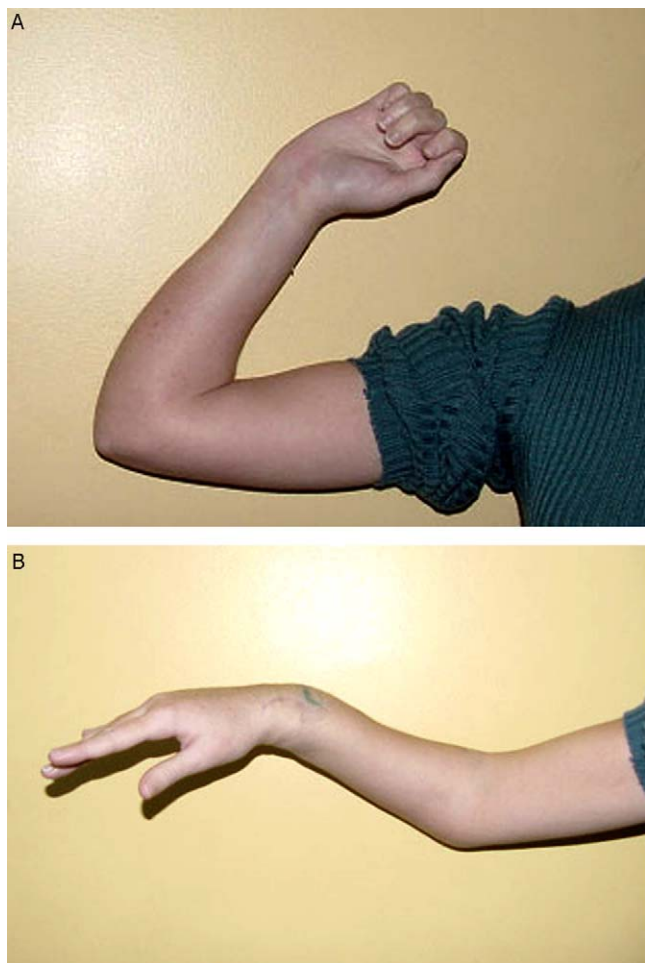


Fig. 3. A, B. Flexion–extension of the elbow and function of the transfer at 3 years follow-up.

two patients who had a positive history of surgery and in whom the tumor had arisen. Rock et al. [3] noticed that 19% of the patients in their series reported a definite episode of trauma, including surgery. In our patient, a relationship between the radial head resection surgery and the tumor growth was suspected.

Desmoid tumors are defined generally as benign tumors by many authors [1,9–12]. Conversely, Posner et al. [13] recognized them as low-grade soft-tissue sarcomas. In any of these situations, the pathology has a marked tendency for local invasion [12,13]. Due to this local aggressive behaviour, most authors recommend surgical excision with wide margins whenever possible [1–3]. The wide resections usually necessary and the high grade of recurrence could leave an important dysfunction of the affected area.

The optimum management and long-term outcome for patients with extremity desmoids remain unclear. Chew et al. [14] described poor results after both radiotherapy and surgery, 41 out of 78 patients had recurrence of the tumor after surgery and ten out of 16 after radiotherapy and surgery. Nuyttens et al. [15] reported a review of 22 published series of desmoid tumors including 780 patients. They concluded that local control of desmoids was better after radiotherapy or radiotherapy with surgery than surgery alone. Pritchard et al. [2] reported 32% of

recurrence without describing anatomic location, while Reitamo [12] noticed 50% of recurrence in the shoulder and upper limb. Other authors reported the recurrence rates between 36 and 68% [1,3].

Several articles reported the functional results of the surgery [16–18], but only three [19–21] described a reconstructive procedure at the same time of the surgery. Ferraresi et al. [19] described an aggressive fibromatosis of the radial nerve which required tumor removal and nerve graft repair, with an excellent clinical recovery and no recurrence at the 6-year follow-up review. Pruzansky et al. [20] presented a case in which a large extra-abdominal desmoid tumor was removed along with the entire deltoid and three-quarters of the triceps and a *latissimus dorsi* musculocutaneous flap was used to replace the resected triceps and deltoid muscle and provide coverage in this region. Goubier et al. [21] reported that one patient out of seven with desmoid tumors located near the brachial plexus that was treated with resection of the tumor that involved the median nerve and a nerve graft was necessary as reconstructive surgery.

In our case, the resection included all the structures of the posterior compartment of the forearm along with the posterior interosseous nerve. This could have left an important dysfunction if we had not performed the reconstructive surgery that included the transfer for the radial palsy, the arthrolysis of the elbow joint and the Sauvé-Kapandji procedure. These reconstructive procedures were useful to recover an acceptable function of the upper extremity.

Although the “oncological” surgery is the most important aspect of the treatment, it is also essential to consider the reconstructive aspect of the surgery when such tumors affect anatomical structures that compromise functions of daily life activities. Furthermore, it is recommended to perform the reconstructive surgery at the same procedure of resection whenever possible.

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