CASE REPORT

Ewing’s sarcoma of the finger: Report of two cases and literature review

S. Baccari, M.F. Hamdi, Z. Mabrouki, M. Daghfous, L. Tarhouni

Department of Hand and Reconstructive Surgery, Kassab Institute of Traumatic and orthopaedic surgery, Manouba 2010, Tunisia
Department of Trauma and Orthopaedic Surgery, Fattouma Bourguiba University hospital, Monastir 5000, Tunisia

Accepted: 20 June 2011

KEYWORDS
Chemotherapy; Ewing’s sarcoma; Finger; Surgery

Summary  Ewing’s sarcoma of the finger is extremely rare. Pain and swelling of the affected finger are the most frequent presenting features. We report two cases of Ewing’s sarcoma located at ring finger and the thumb in two children aged 14 and 10 years. The first patient died of generalised metastases despite surgery, chemotherapy and radiation therapy. The second had no localised recurrence or metastases after surgery and chemotherapy at last follow-up of 4.5 years after tumour resection. The tumour’s surgical accessibility, chemotherapy, and radiation therapy improve the prognosis of this tumour.

Introduction

Ewing’s sarcoma was first described in 1921 [1,2]. It is the second most frequent primary bone cancer of childhood and adolescence, after osteosarcoma [3]. It usually affects the long bones, but in the hand this tumour is rare, and exceptional in the finger. Different treatments using surgery, chemotherapy, radiotherapy, and their combinations are described. The aim is to save the hand and the patient’s life. We report two cases of Ewing’s sarcoma, one in the thumb and the other in the ring finger.

Cases report

Case report 1

A 14-year-old, right hand dominant boy presented with 11 months history of a gradual pain and tumefaction involving the proximal part of the left ring finger (Fig. 1). There was no recent trauma or fever or loss of weight. Locally, the swelling was indurate; the range of motion of the metacarpophalangeal joint was restricted.

The X-ray examination of the affected finger, revealed bone destruction with sclerosis and soft tissue component of the proximal phalanx (Fig. 2). The laboratory findings were within normal limits. At the Magnetic Resonance Imaging (MRI), the tumour was detected with more precision (Fig. 3); and the tumour appeared to be malignant on MRI.

Open surgical biopsy was performed. The histology confirmed the diagnosis of Ewing’s sarcoma. The preoperative
assessment including chest tomography, and whole-body bone scan showed no metastatic disease. Preoperative chemotherapy (VIDE protocol) was indicated and used for six courses using vincristine, ifosfamide, adriamycin, ectoposide. After the parent’s consent, a wide excision of the tumour and the involved tissues (ray amputation) was carried out without local reconstruction (Fig. 4). The surgical margins were negative. The histological findings proved a poor response to chemotherapy. After surgery, a second treatment course of chemotherapy using vincristine, actinomycin D, ifosfamide, was administered for six courses. The radiotherapy was indicated because the histological study of the excised tumour revealed more than 10% of tumour cells; the area of the tumour was irradiated with 40 Gy. Six months later, the patient developed metastases in the lung and tibia. Unfortunately, the patient died of generalised metastases 41 months after the diagnosis of the tumour was established.

Case report 2

A 10-year-old, left hand dominant boy presented with a mass arising in the distal phalanx of the left thumb for 4 months. This isolated mass was fixed and painful (Fig. 5). The radiological examination revealed permeative bone destruction and perpendicular spicules of the distal phalanx of the left thumb (Fig. 6). A biopsy was performed.
The histopathological diagnosis was Ewing’s sarcoma. The chest tomography showed no metastatic lesions. A technetium bone scan revealed increased uptake only in the distal area of the left thumb. Amputation at the metacarpophalangeal level was carried out (Fig. 7). The patient had postoperative adjuvant chemotherapy using vincristine, ifosfamide, adriamycin, etoposide for six cycles. The patient did not develop metastasis or local recurrence; he was well and alive at 4.5 years after surgery.

Discussion

Initially Ewing’s sarcoma was described as a malignant tumour of bone origin, invading the soft tissue. The origin of Ewing’s sarcoma is a subject of much debate. Once thought to be derived from primitive neuroectodermal cells [4], many now believe it to arise from a mesenchymal stem cell [5]. We must mention Ewing tumour and not Ewing sarcoma [4].

Genetic factors play a role in its pathogenesis. There is a form of anomaly chromosomal translocation of two chromosomes 22 and 15 [6]. Usually this tumour occurs during the first or second decade of life; it particularly affects long bones and pelvis [7]. In the hand Ewing sarcoma is very rare, Kissane JM series of 303 cases of Ewing sarcoma, identified one lesion in the hand [8]. The metacarpals are most commonly affected and occurrence at the phalanges is quite rare [9–11]. Regardless of the treatment modality, the location of the tumour in the hand is an important factor of prognosis [10]. Because the survival is highest in lesions of the distal bones of the extremities [8].

The review of literature found 15 cases of Ewing’s sarcoma of the finger [1,3,9–17]. We report the summary of those cases (Table 1). According to literature review, males were more affected (69%); the average age of detection of the tumour was 18.5 years (5 months–51 years). The tumour was located at the proximal phalanx in 53% of cases. The thumb (28%) and the long finger (28%) were the most affected fingers.

Pain and swelling of the affected fingers were the most frequent complaints at diagnosis [7], initially the patient had a general good health, and there were no fever and loss of weight [4]. Sometimes the tumour is clinically mistaken for a local infection because of pain and local inflammatory symptoms, X-ray findings can also be mistaken for osteomyelitis because of the bone destruction [1].

Several studies [4,7,9,10,17] have suggested wide radical resection of the tumour, combined with chemotherapy as the best therapy. This is easily accomplished in case of digital localisation. The surgical accessibility of Ewing’s sarcoma of the phalanx makes them amenable to radical resection [1,11]. Two cases of Ewing’s sarcoma of the thumb [11,14] were treated by primary amputation without radiation therapy or chemotherapy. There was no evidence of disease in 26 to 60 months following up.

Figure 5  Clinical picture of the multinodular tumour with necrotic area involving the distal part of the left thumb.

Figure 6  An X-ray film of the left thumb showing bone destruction with perpendicular spicules of the distal phalanx. Soft tissue mass visualized in the border of distal phalanx.

Figure 7  No local recurrence of the tumour after metacarpophalangeal disarticulation.
Radiotherapy alone is not indicated because it cannot control the primary lesion, and induces major physical damage (soft tissue contracture, growth disturbances in young patient, secondary radiation-induced malignant tumour) [4,9]. The radiation is beneficial when wide margins cannot be surgically achieved or when the response to chemotherapy is incomplete [3].

In spite of the adjuvant therapy, most authors agree about the benefit from the tumour’s resection, which remains the most important stage of the management of Ewing’s sarcoma of the finger. Kinsella [18] suggested the combination of chemotherapy with radiotherapy without surgery, in controlling the primary lesion in the hand and to avoid a hand mutilating surgery, the long-term result remains uncertain. Ewing’s tumours of the bones are more susceptible to chemotherapy [19]. This therapy is an integral part of the treatment in preoperative and postoperative surgery of the primary tumour, adjuvant chemotherapy adapted to the histological response to the neoadjuvant treatment.

The treatment of Ewing’s sarcoma is achieved through a Common European protocol called Euro Ewing 99 [20]. The prognosis depends on the tumour volume and histological response to initial chemotherapy type VIDE which associated vincristine, ifosfamide, Adriamycin, and Etoposide. After surgery, the chemotherapy was administered. The tumours will be irradiated if the histological study revealed more than 10% of tumour cells after surgery and chemotherapy [4].

The final surgical management is to achieve an acceptable cosmetic appearance and excellent function of the hand. After the amputation of a finger, the reconstruction is possible for aesthetic and functional reasons. The reconstruction was achieved at the same time of tumour resection [9], or remotely after tumour excision [4].

Despite the extremely rarity of this tumour finger arising in the finger, the diagnosis should be considered in any lesion suspicious for malignancy. The treatment is urgent and specialized. Currently, the most favourable treatment of Ewing’s sarcoma of the finger combines surgery and chemotherapy. The histological findings determine the usefulness of radiotherapy. Despite the small size of these tumours the preoperative chemotherapy is necessary.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

**References**

Ewing’s sarcoma of the finger: Report of two cases and literature review