Plexiform Histiocytic Tumor of the Thumb

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Abstract

Introduction

The plexiform fibrohistiocytic tumor is a conjunctive intermediate malignancy tumor. The related literature is poor. It is usually encountered in female children and adolescents. Its symptomatology is also poor. The clinical symptomatology of this disease is sometimes marked by rare dermal or subcutaneous multinodular lesions. They evolve slowly. The diagnosis must be made early to perform wide surgical excision. This tumor is characterized by the risk of local recurrence.

Objective

The aim of this work was to describe an unusual clinical form by its onset predisposition and its symptomatology at an advanced stage.

Our Observations

Our patient was a 58-year-old male with a periungal swelling of the right thumb. This swelling had required unsuccessful multiple surgical cures. At our consultation 5 months after the last cure, this swelling presented itself in the form of a budding cauliflower tumor. The histological examination of the specimen led to the diagnosis of plexiform fibrohistiocytic tumor. Proximal transphalangeal amputation was then performed. At the 14-month follow-up, the progression was simple and the outcome was normal.

Conclusion

Our observation shows that the plexiform fibrocystic fibrosis tumor which occurs more often in the female child can also be discovered in the male adult. At a later stage its diagnosis must be suggested in the presence of a budding cauliflower tumor.

Keywords: Plexiform Fibrohistiocytic Tumor; Soft Tissue Sarcomas

Introduction

Plexiform fibrohistiocytic tumor (PFHT) is an intermediate malignancy tumor recently described [1]. It is rare and affects mainly the child and young adult. This tumor is the most reported in adolescents with an average age of 15 years. The seats of predilection are the upper limbs. It is generally asymptomatic and develops slowly. The clinical symptomatology of this disease is sometimes marked by rare dermal or subcutaneous multinodular lesions. These are usually less than 3 cm in diameter. They evolve slowly. The recurrence rate is 40% [2,3,4]. We report a case that is characterized by its epidemiological and evolutionary features.

Observation

A 58-year-old male consulted for a budding swelling of the right thumb. This swelling was of gradual appearance, invading the nail and had been developing for 4 years. There was no evidence of trauma or hand irradiation in his history. There was no notion of prolonged drug intake. Three excisions had been performed and led to the diagnosis of non specific sarcoma of the soft tissue. The general condition of the patient was normal. At the consultation carried out 5 months after the last excision, there was a budding tumor with a “cauliflower” appearance (Figure 1). A bone invasion in the form of osteolysis has been shown on the X-Rays (Figure 2).

Figure 1: budding “cauliflower” Appearance of histiocytic tumor that has been developing for 2 years.

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We performed a proximal transphalangeal amputation of the thumb. The histology of the specimen concluded that it was plexiform fibrohistiocytic sarcoma of intermediary malignancy. It was about dermal and hypodermic nodules with giant epithelioid, fusiform cells of the osteoclastic type in chronic inflammatory infiltrate (Figure 3). After healing, we installed a polyethylene prosthesis. At the 14-month follow-up, the patient's general condition was retained. There was no local recurrence (Figure 4). Extension assessment with standard chest X-ray, abdominal ultrasound and abdominal CT were normal. After this consultation we proposed the thumb reconstruction by toe transfer [5]. After an interview with the patient, this latter refused this intervention because of the inadequacy of the technical platform, the cost, and the risks of failure of this microsurgery.

**Discussion**

Our case is unique in several respects. This disease, usually described in the female child, was observed in a 58-year-old male patient. These are generally asymptomatic lesions, evolving slowly. Generally, the authors describe this tumor at the stage of dermal or sub-dermal nodules of less than 3 cm in diameter [3,6]. At this stage the diagnosis must be accurate in order to propose a suitable treatment based on surgical excision. This diagnosis is based on clinic and histology. The aim of the treatment is to achieve the most complete excision possible [1,2]. No chemical or radiotherapy treatment has yet been published. Our observation describes a highly developed tumor that could raise a differential diagnosis problem. Notably with the ingrown nail because of the frequency of the recurrence. At an advanced stage the diagnosis can be made with the spinocellular epithelioma of Marjolin ulcers [7] which also give a budding appearance in cauliflower. Thus at the stage of our patient, the clinical examination no longer allowed to direct the diagnosis. This diagnosis was assured by the histology.
Figure 5: Postoperative aspect at 14 months of the surgical specimen. The amputation was proposed right away because we took into account the previous diagnosis of soft tissue sarcoma, the presence of bone lysis, the notion of recurrence and the exaggerated volume of the tumor. The evolution of the Plexiform fibrohistiocytic tumor even after early and massive excision is often accompanied by recurrence [1,2,8]. No recurrence has been observed in our patient, but the short follow-up does not allow to assess the long-term evolution of the disease which requires a rigorous and continuous monitoring.

Conclusion

Our observation points out that the plexiform fibrohistiocytic tumor which occurs more often in the female child can also be discovered in the male adult. At a later stage its diagnosis must be suggested in the presence of a budding cauliflower tumor.

Author’s contribution

This study was approved by the local ethics committee and all the authors contributed to the writing of this.

References