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Desmoid Tumor in Hand: A Case Report

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Abstract

The desmoids tumors are characterized by the excessive uncontrolled proliferation of connective tissue. Although benign, it has an aggressive nature that invades and permeates surrounding tissues and organs, resulting in the destruction, loss of function and deformity of the adjacent structures. It is a rare tumor, only 0.03% of all cancers, and whose etiology is still unknown. In this report, we describe a patient who has developed a desmoid tumor in her hand and discuss the treatment established.

Introduction

The Desmoid Tumor (DT) is a tumor characterized by local and aggressive proliferation of the connective tissue that infiltrates and invades organs and neighboring structures, especially fascias and muscles [1]. This pattern of development is also present in others fibroproliferative's diseases, such as the Dupuytren's Disease (DD), which leads to the thickening and retraction of the palmar fascia and digital extension deficit due to excessive amount of collagen type 3 [2]. From the anatomopathologic perspective, the lesion is described by well differentiated fibroblasts interlaced with collagen bundles and cellularity varies. There is no capsule and it is a benign tumor with low metastases potential [3].

However, it has a high recurrence rate when approached surgically: for primary tumors, in the first 5 years, the number of relapse is about 80 to 90%. After this period, it reduces to 56% depending on the size, location, patient age and size of the resected margin [4]. Besides that, the rapid growth results in local destruction, loss of function and deformity of the adjacent structures.

They are rare tumors representing 0.03% of all neoplasias and less than 3% of all soft tissue tumors [5]. They mainly affect young people between 15 and 60 years of age, with a peak age of about 30, and are discreetly more common in women [6]. The DT's etiology is still unknown. However, there are several hypotheses explaining the appearance of DT, for example, the occurrence of local trauma or body's reaction to the surgical incision. At the molecular level, some genes mutations have been detected: modifications in beta-catenin genes were found in 85% of sporadic DT, which can also be associated with Familial Adenomatous Polyposis (FAP), since about 80% of patients who developed intra-abdominal DT had had also FAP [6]. A possible explanation for this association is the occurrence of molecular transformations, during the healing process, and fibroproliferative disorders in the mesenchymal tissue [7].

Case Report

Female patient, 25 years old, searched for care assistance because of the presence of grease on the anterior side of the right hand. The tumor was adhered to deep planes and painful to the touch.

The patient reports that the injury began about one year ago, when it was characterized by a small and painful node that appeared after a blunt trauma. Since then, the lesion presented progressive increase in size. During the physical examination, it was possible to see the local commitment of her hand and a severe restriction on the full flexion of the metacarpal phalangeal joints observed in fingers II, III and IV. There was no change in sensitivity (Figure 1).

The MRI's patient showed multiples and focal intrasubstance lesions involving fatty muscles plans and ligaments structures of the anterior and posterior surface of the right hand. Furthermore, the majority part of the lesion affects the hypothenar eminence reaching the metacarpal phalangeal joint structures and from the second to fourth proximal interphalangeal (Figure 2).

In the case reported in this article, it was necessary to carry out biopsy and pathology test which revealed the presence of fibrous tissue by fibroblasts proliferation between dense collagen stroma (HE staining). However, histopathology was not sufficient for final diagnosis, since the characteristics presented resemble various diseases, for example, the DD. Therefore, in order to deepen the case study, immunohistochemical was performed. The method comprises the blade material reacting with specific antibodies (staining for DNA analysis) in order to determine the site of each protein. At the end, it was searched for the estrogen receptors (6F11), progesterone

Figure 1: Posterior and anterior view of patient's hand.

receptors (PsR320), S100 protein, CD34 receptors, marker presents in mesenchymal and fibrous tumors, Vimentin (3B4) and B-catenin which are closely correlated with fibroproliferative dysfunction. The result was negative to estrogen receptors (6F11), progesterone (PsR320) and S100, and it was positive for vimentin (3B4), CD34 and Beta-catenin (Figure 3 and Figure 4).

Discussion

The fibromatosis diseases can be divided into two groups: superficial or deep. The first one has three main classes: the DD, with palmar fascia involvement, the Ledderhose Disease (LD), plantar fascia, and finally the Peyronie Disease (PD), penile fascia. The second group corresponds to the TD, which can be intra-abdominal, abdominal or extra-abdominal. Therefore, the determination of the right diagnosis is extremely important to choose the best treatment conduct in each case [8].

Although benign, it has an aggressive nature and an invasive behavior, resulting in the destruction and deformity of the structures, which reduces the functionality of patient's hand, since there is a significant damage to the plans involved and important structures as well.

Because it is an extensive and profound injury, it requires a differentiated and effective therapeutic approach, once, in this case, the surgery presents mutilate character and it can't be considered to treat

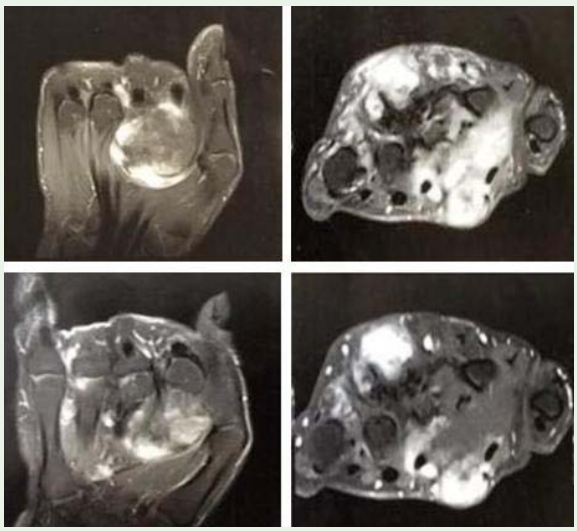

Figure 2: Axial and sagittal cuts of RMI's patient showing multiples lesions.

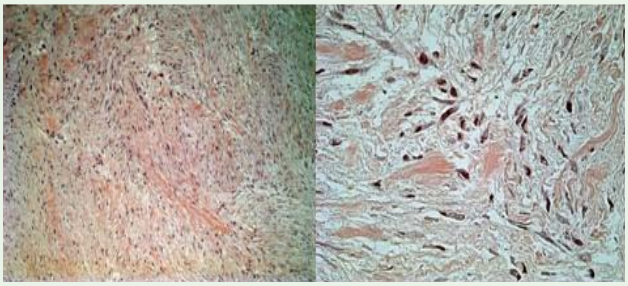

Figure 3: Histopathology showing fibrous tissue into dense collagen, HE staining, 400X.

this type of tumor [6]. Therefore, due to the impossibility of complete resection of the lesion, the conduct adopted was chemotherapy following the drug regimen: doxorubicin and dacarbazine in 5-6 cycles every 21 days.

Currently, there is a proposal known as "wait and see", which suggest following the tumor's evolution without direct interference by drugs, radiological or surgical procedures. Studies show that 85% of tumors underwent spontaneous reduction, after 1 year of evolution, and half of the DT had their size stabilized to that period [8]. These results show great effectiveness in the application of this method, especially in primary DT and recurrent extra-abdominals types [9]. All this in order to ensure the preservation of member function and to reduce the recurrence rates evidenced in the surgical and radiotherapy procedures [10].

Conclusion

The patient's DT, although benign, has an aggressive nature that invades the tissues, resulting in the destruction, loss of function and deformity of the adjacent structures. All this compromises the global dynamic of the member. The traditional treatment is based, primarily, on surgical resection plus adjuvant radiotherapy. However, in many cases, surgery can't be performed due to the large local involvement. Therefore, the doctors try different conducts: the chemotherapeutic treatment, drugs treatments by anti-inflammatory and hormonal therapy [4].

In the case presented, it was decided to treat with chemotherapy, since the surgery and radiotherapy are contraindicated and there was no prior response to the treatments with anti-inflammatory drugs or hormonal therapy.

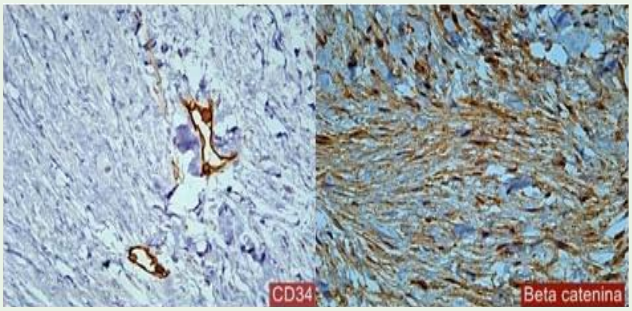

Figure 4: Immunohistochemical specific staining detecting CD34 receptors and Beta-catenin, positive in cases of DT.

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