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Case Report

Aggressive fibromatosis of head and neck: Considerations on the optimal treatment method on the basis of case report

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ABSTRACT

Introduction: The term fibromatosis refers to proliferation of fibrous tissue in the form of irregular nodules or soft tissue sarcoma. It is characterized by local malignancy with tendency to recurrence, significant hypertrophy of the mature connective tissue and lack of the ability to form distant metastases. Pathological lesions originate from fascial, muscle and tendinous tissue.
Aim: The aim of this work is to analyze the efficacy and safety of the selected methods of treatment of aggressive fibromatosis on the basis of the presented case.
Case study: A five-year-old boy was admitted to the Department of Otolaryngology of the Provincial Specialist Children's Hospital in Olsztyn due to right sided cervical mass. Histological examination of biopsy material revealed aggressive fibromatosis. The child was previously treated for two years with systemic chemotherapy with methotrexate, vinblastine, vincristine, imatinib and tamoxifen with no improvement. Surgical tumor removal was performed on February 14, 2014. During the 18-month observation period no signs of tumor recurrence were observed.
Results and discussion: In the treatment of aggressive fibromatosis various therapeutic methods are used, including radiotherapy, chemotherapy and surgery. The effects of chemotherapy and radiotherapy are frequently unsatisfying. These methods are associated with numerous complications and side effects, thus mode of treatment should be considered individually for each patient. In case of the presented patient surgical tumor removal was the best and the most effective treatment method.
Conclusions: Surgery should be a treatment of choice, whereas chemotherapy and radiotherapy should be considered as alternative or complementary methods in the treatment of aggressive fibromatosis.

Introduction

The term fibromatosis refers to a wide group of fibroplastic lesions which present as low grade soft tissue sarcoma. They are characterized by local aggressiveness with a tendency to local recurrence, significant hypertrophy of mature connective tissue with formation of conglomerate of nodules and lack of the ability to form distant metastases. In English literature it is known as aggressive fibromatosis (AF) or desmoid tumor, distinguished by deep location.[1]

Histologically, typical features of fibromatosis include excessive proliferation of homogeneous slender spindle cells infiltrating the adjacent tissue. They are surrounded by collagen stroma with visible vessels. These cells show no atypia and their mitotic index may vary within broad limits. Number of mitoses may be focally high, which does not imply malignancy. Pathological lesions originate from fascial, muscle and tendinous tissue.[1,2]

Correlation between injury and AF was revealed in 19%–49% of cases. There is a theory that its pathogenesis is associated with an abnormal wound healing process with proliferation of immature fibroblasts, which subsequently leads to tumor formation. The role of sex hormones in pathogenesis of AF is also emphasized.[2,3]

Depending on the location of lesion fibromatosis was divided into two clinical types: superficial and deep. The most common type of superficial fibromatosis is palmar fibromatosis, called Dupuytren's contracture. Other typical locations of superficial AF

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include feet and penis.[4] Deep fibromatosis most frequently locates in the abdominal wall, typically in straight abdominal or internal oblique muscle. Deep AF constitutes approximately 3% of soft tissue tumors, and there are 2–4 new cases per 1 million people each year. Intraabdominal form locates in the mesentery and pelvis minor. Mesenteric fibromatosis is associated with Gardner's syndrome.[5]

Typical extra-abdominal locations include shoulder, chest wall, back, thigh, head and neck. In case of extra-abdominal locations predominant symptom is poorly demarcated deep-seated tumor. Other symptoms include edema and pain associated with infiltration of the adjacent structures and organs. Probability of the occurrence of AF of the head and neck is almost equal for both sexes. Although AF does not tend to metastasize, in case of head and neck location course of the disease may be unfavorable and lead to serious complications, including death.[6–8]

2. Aim

The aim of this work was to analyze the efficacy and safety of the selected methods of treatment of AF on the basis of the presented case.

3. Case study

Male child aged 5 years 4 months was admitted to the Department of Otolaryngology of the Provincial Specialist Children's Hospital in Olsztyn due to right sided cervical mass for surgical treatment. At the age of 2.5 years he was diagnosed with right sided cervical tumor which presented after upper respiratory tract infection. Magnetic resonance imaging (MRI) of the neck conducted in September 2011 revealed an abnormal tissue mass with irregular borders in the right submandibular area. The dimension of the tumor was $5.3 \times 2.9 \times 4.4$ cm. The lesion was posteriorly directly adjacent to cervical vessels, not infiltrating them, while medially it reached parapharyngeal space (Fig. 1).

Material collected during aspiration biopsy revealed nodular fasciitis. In January 2012 open biopsy of the tumor was performed and diagnosis of AF was established (report no. 8352/2012 dated February 8, 2012, Department of Pathology, Medical University of Wroclaw). In November 2012 chemotherapy with methotrexate and vinblastine was initiated, and overall 10 courses were administered. The treatment was completed in January 2013. No clinical improvement was observed and MRI revealed slight progression of the tumor.

In March 2013 another chemotherapy was started. Vincristine was used but after the third course the treatment was discontinued due to gastrointestinal complications. From May 2013 patient was treated with imatinib (Glivec) and from November 2013 with tamoxifen. After tamoxifen slight regression of the tumor was observed, confirmed in MRI of the neck. Due to unsatisfying effects of conservative treatment patient was qualified for surgery (Fig. 2).

Surgical tumor removal was performed on February 14, 2014 (Prof. A. Kukwa). The procedure involved incision along the sterocleidomastoid muscle, dissection entirely within healthy tissue of the tumor penetrating in the region of large cervical vessels and lateral pharyngeal wall. Entire submandibular gland was also removed. During postoperative period House-Brackman Grade III paresis of the lower branch of the facial nerve and difficulty swallowing solid foods were observed within first days after surgery and resolved spontaneously. Patient was discharged home in good general condition on the 12th postoperative day and remains in follow-up (Fig. 3).

Histological examination (report no. K91/14 dated March 17, 2014, Department of Pathology, Institute of Mother and Child in Warsaw) confirmed previous diagnosis of AF (*fibromatosis* p. *desmoides*) (Figs. 4 and 5).

Control MRI of the neck after surgery was performed three times, last in June 2015 and showed no tumor recurrence (Fig. 6).

4. Results and discussion

In the management of AF surgical removal of the tumor with a margin of healthy tissue is a treatment of choice. However, it is not always possible due to tumor sizes, penetration to adjacent structures and tissues, as well as macroscopically unclear margin of the tumor. Local recurrences after AF resection in the head and neck region are estimated at 40%–70%. The majority of recurrences are observed within 18 months post surgery.[9]

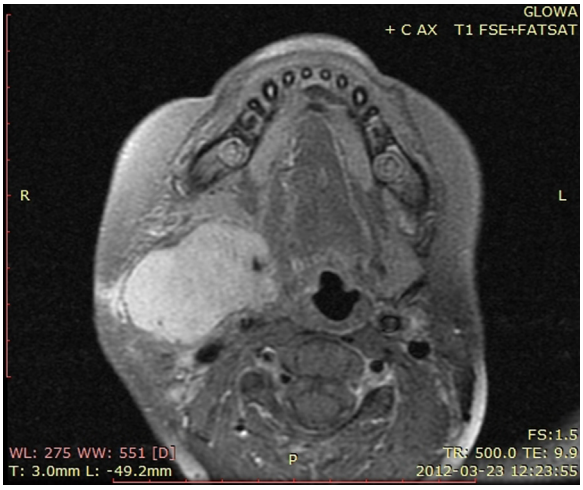

Fig. 1. MRI of the neck before surgical treatment. Visible large right sided tumor enhanced after contrast infusion.

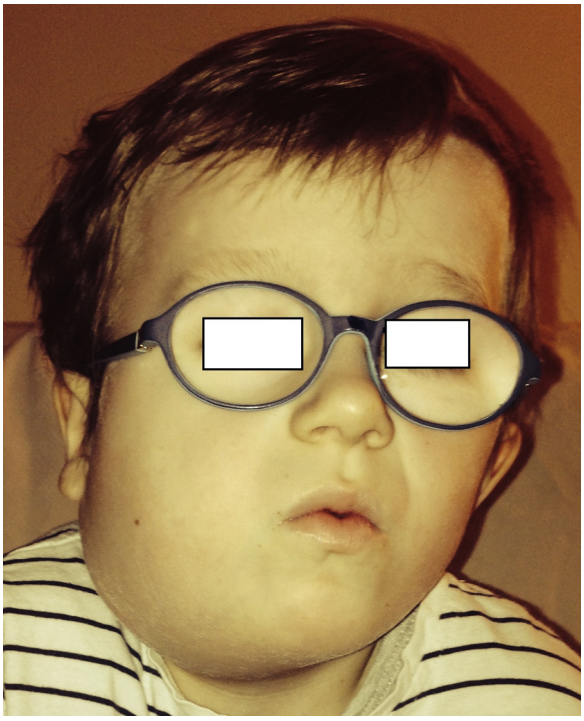

Fig. 2. Patient before surgery. Visible large asymmetry of the right neck caused by the presence of the tumor.