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DESMOID-TYPE FIBROMATOSIS OF THE NECK WITH BRACHIAL PLEXUS INVOLVEMENT: A RARE CASE REPORT

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ABSTRACT

Introduction: Desmoid-type fibromatosis (DTF) is a rare soft tissue tumor arising from musculoaponeurotic connective tissue, characterized by locally aggressive growth and a high rate of local recurrence despite the absence of metastatic potential.

Case summary: We report a case of a 19-year-old female patient presenting with a right cervical–supraclavicular mass associated with progressive upper limb numbness. Magnetic resonance imaging showed a large soft tissue mass infiltrating adjacent muscles and surrounding the bundles of the brachial plexus. Ultrasound-guided needle biopsy confirmed the diagnosis of desmoid-type fibromatosis. The patient underwent surgical dissection with maximal tumor resection combined with decompression and preservation of the brachial plexus. Postoperative histopathological results were consistent with DTF, with immunohistochemical staining for β-catenin positive. The postoperative course was favorable, with marked improvement in neurological symptoms and preservation of upper limb function. Although postoperative magnetic resonance imaging revealed residual lesions, the patient was selected for periodic follow-up instead of radiotherapy due to the risk of long-term radiation-related complications in a young patient.

Conclusion: This case demonstrates that maximal tumor resection combined with preservation of neural structures may be an appropriate treatment strategy for desmoid-type fibromatosis of the cervical–supraclavicular region involving the brachial plexus.

Keywords: *Desmoid-type fibromatosis; desmoid tumor; brachial plexus.*

1. INTRODUCTION

Desmoid-type fibromatosis (DTF), also known as desmoid tumor or aggressive fibromatosis, is a rare tumor arising from fibroblastic cells of musculoaponeurotic connective tissue. Although it is classified as a benign tumor due to the absence of metastatic potential, it is characterized by locally infiltrative growth and a high rate of recurrence after treatment, thereby posing significant challenges in clinical management [1].

DTF is a rare condition, accounting for approximately 0.03% of all tumors and less than 3% of soft tissue tumors, with an estimated incidence of 2–4 cases per million population per year. The disease most commonly occurs in individuals aged 15 to 60 years and shows a female predominance [2]. Based on anatomical location, DTF is classified into three main groups: extra-abdominal, abdominal wall, and intra-abdominal, of which extra-abdominal tumors account for the highest proportion. The occurrence of tumors in less common locations, such as the chest wall or the head and neck region, is relatively rare.

The pathogenesis of DTF remains incompletely understood. Several studies have shown that the disease is associated with dysregulation of the Wnt/β-catenin signaling pathway, in which mutations in the CTNNB1 gene are commonly observed in sporadic cases, while mutations in the APC gene are more frequently found in patients with familial adenomatous polyposis [3]. In addition, several other risk factors have been reported, including local trauma, a history of surgery, and hormonal factors; however, the exact role of these factors remains controversial.

Clinically, DTF typically presents as a slowly growing soft-tissue mass located deep within the soft tissues and often associated with minimal pain. However, due to its locally infiltrative nature, the tumor may extend into adjacent structures, such as muscles, blood vessels, or nerves, thereby causing functional symptoms depending on the lesion's anatomical location. Imaging modalities, particularly magnetic resonance imaging (MRI), play an important role in evaluating tumor size, extent of infiltration, and its relationship with surrounding anatomical structures. The definitive diagnosis is based on histopathological examination and immunohistochemical staining for β-catenin, which helps differentiate DTF from other soft-tissue spindle cell tumors.

The management of DTF remains controversial and often requires a multimodal and individualized approach for each patient. Treatment options may include active surveillance, surgical resection, radiotherapy, or systemic therapy, depending on tumor location, size, and clinical symptoms [4]. Among these, surgery remains a commonly selected treatment modality for symptomatic or progressive tumors, although the rate of local recurrence remains relatively high.

We report a case of desmoid-type fibromatosis of the right cervical–supraclavicular region with symptoms caused by tumor infiltration of the brachial plexus in a young female patient, treated with maximal tumor resection and nerve preservation, in order to contribute to clarifying the challenges in the diagnosis and management of this rare condition.

2. CASE REPORT

A 19-year-old female patient, previously healthy, with no history of local trauma, prior surgery, or family history of soft tissue tumors,

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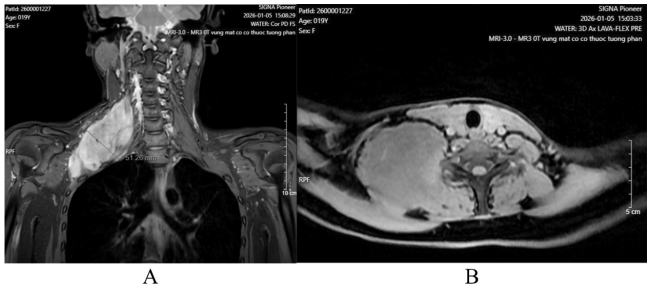
presented with pain and numbness of the right forearm and hand for approximately 2 months. On clinical evaluation, a mass was detected in the right cervical region. Over the past 3–4 months, the mass had increased rapidly in size, accompanied by worsening numbness of the right hand, without any motor weakness.

On physical examination, the right cervical region was more prominent than the left, with a palpable mass in the right cervical–supraclavicular region, firm in consistency, non-mobile, and non-tender on palpation. The right upper limb was associated with numbness and pain along the anteromedial aspect, radiating to the little finger, with gradually increasing severity over time. Contrast-enhanced magnetic resonance imaging demonstrated a soft tissue mass in the right cervical and supra- and infraclavicular regions, as well as the right chest wall, measuring 95 × 43 × 60 mm, with irregular margins and indistinct boundaries from the surrounding soft tissues (Figure 1). The lesion infiltrated adjacent muscles and surrounded the brachial plexus bundles, resulting in loss of normal anatomical delineation of these neural structures. However, no evidence of invasion into adjacent bones was observed. Ultrasound-guided core needle biopsy showed fibromatosis of connective tissue origin.

The patient underwent surgical adhesiolysis, near-total tumor resection, and decompression of the right brachial plexus. Intraoperatively, a firm mass was observed in the right cervical region, infiltrating and adherent to the surrounding soft tissues. The tumor extended from the anterior border of the right sternocleidomastoid muscle posteriorly, where it was tightly adherent to the spine. It displaced and deformed the brachial plexus anteriorly and medially, with part of the tumor encasing the nerve roots. Inferiorly, the tumor compressed the subclavian vein. The tumor was maximally resected while preserving the brachial plexus and vascular structures. Histopathological examination was consistent with desmoid fibromatosis infiltrating the muscle and surrounding fibro-fatty tissue. Tumor cells showed positive immunohistochemical staining for β-catenin, which supported the diagnosis of desmoid-type fibromatosis and helped differentiate it from other spindle-cell soft tissue tumors. In addition, activation of the Wnt/β-catenin signalling pathway has been associated with the locally aggressive behaviour and high risk of local recurrence observed in desmoid tumors. (Figure 2)

The postoperative course was favorable, and the patient recovered well and was discharged after 7 days of treatment. At the 1-month follow-up, the patient reported marked improvement in clinical symptoms, with complete resolution of numbness, and normal motor and sensory function of the right upper limb. (Figure 3) Follow-up MRI after surgery showed a residual soft tissue lesion in the right supra- and infraclavicular region and chest wall, measuring approximately 31 × 41 mm, suggestive of residual tumor after surgery. (Figure 4) The patient and her family were fully informed about the disease characteristics, the risk of recurrence, and the long-term follow-up plan. The patient was advised to undergo regular follow-up with clinical and imaging evaluation.



A B

Figure 1. Preoperative magnetic resonance imaging of the cervical–thoracic region (A: sagittal view, B: axial view)

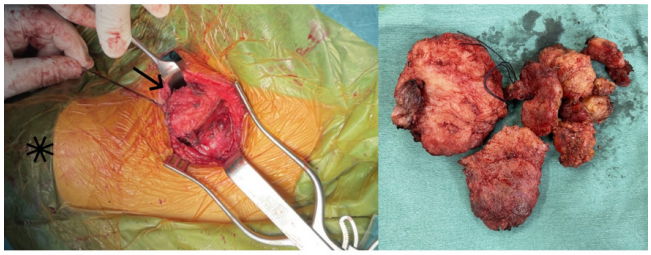


Figure 2. A: Intraoperative image (*: right shoulder, arrow: tumor), B: Tumor specimen

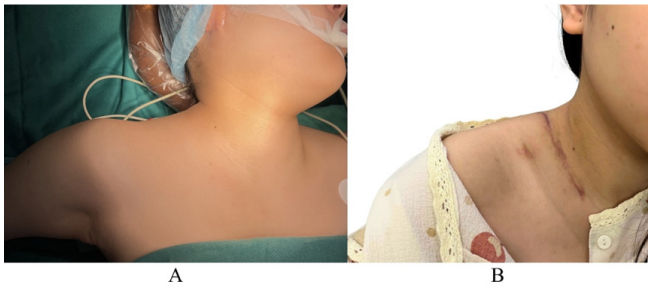


Figure 3. A: Preoperative image; B: Postoperative image

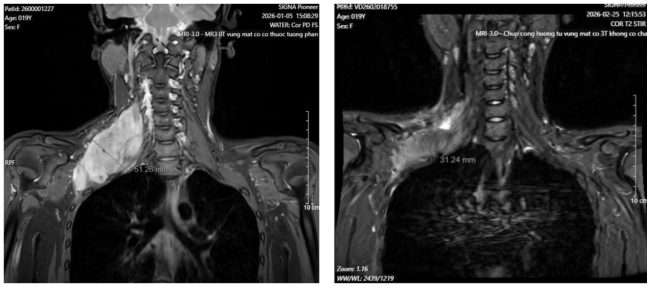


Figure 4. Sagittal magnetic resonance imaging (A: Preoperative; B: Postoperative)

3. DISCUSSION

Desmoid fibromatosis is a rare benign tumor with no metastatic potential, characterized by local infiltration and a high rate of recurrence. According to a review by Bektas M et al., the disease most commonly occurs between the ages of 20 and 44 years, predominantly in females, with a female-to-male ratio of 2–4:1 [5]. The tumor can occur in various locations; however, the cervical–supraclavicular region and upper chest wall are relatively rare sites, accounting for 4–10% of all DT cases [6,7]. Tumors in this region often present significant therapeutic challenges due to the presence of important anatomical structures, including major blood vessels and particularly the brachial plexus. Infiltration or encasement of neural structures may lead to neurological symptoms, such as numbness, pain, or upper-limb weakness. In our case, the patient presented with progressive numbness of the upper limb, accompanied by gradually increasing pain. For the diagnosis of cervical masses, especially when clinical manifestations are suggestive of brachial plexus compression, magnetic resonance imaging is the most appropriate modality. MRI findings in this case demonstrated a tumor encasing and obscuring the boundaries of the brachial plexus bundles. Preoperative biopsy helps confirm the nature of the lesion and exclude other malignant tumors.

The selection of treatment strategy for DTF remains controversial, and there are currently no specific guidelines for choosing the optimal treatment modality. However, two important recommendations widely adopted have been proposed by the National Comprehensive Cancer Network (NCCN) and the Desmoid Tumor Working Group (DTWG) [8,9]. Treatment options include active surveillance, surgery, radiotherapy, chemotherapy, and local therapies [5,8].

- Active surveillance is the first-line approach for tumors that are asymptomatic or mildly symptomatic and non-progressive. During the surveillance period, if the tumor continues to progress or symptoms worsen, subsequent treatment selection depends on the tumor's anatomical location and the extent of its invasion into surrounding structures.

- Surgery is indicated in cases where the

tumor invasion causes severe clinical symptoms, with the goal of maximal functional preservation and a low rate of surgery-related complications..

- Radiotherapy may be administered alone or performed after surgery to achieve better control of local progression as well as recurrence. Adjuvant radiotherapy is recommended in cases with positive surgical margins or in recurrent DTF cases that are difficult to operate on or associated with a high risk of surgery-related complications.

- Systemic therapy, including low-dose chemotherapy, conventional chemotherapy, or tyrosine kinase inhibitors (sorafenib, imatinib, pazopanib, sunitinib), is used in patients with unresectable, rapidly growing, symptomatic tumors or progressive disease. The choice of treatment depends on the extent of tumor invasion and the need for a more rapid therapeutic response [10]. However, systemic therapy may be considered unsuitable due to its potential toxicity, particularly in young female patients, in whom preservation of ovarian function and fertility is of great importance.

In our case, the patient underwent surgical treatment due to tumor progression, rapid increase in size, and the presence of clinical symptoms related to brachial plexus compression. During surgery, important structures were preserved as much as possible, including the brachial plexus and the subclavian vascular bundle, while maximal tumor resection was performed. Previously, wide surgical resection with the goal of achieving negative margins was considered the standard treatment. However, many recent studies have shown that margin status is not always an independent prognostic factor for recurrence, particularly when radical resection requires the sacrifice of important functional structures. Therefore, current treatment trends increasingly prioritize functional preservation and favor maximal tumor resection over radical resection at all costs. In this case, the tumor was closely related to the brachial plexus; therefore, complete tumor resection could lead to a high risk of severe nerve injury and permanent upper limb functional impairment. Accordingly, we selected a strategy of surgical dissection, adhesiolysis, and maximal tumor resection while preserving important neural and vascular structures. After surgery, the patient showed marked improvement in neurological symptoms, and upper limb function was completely preserved.

The recurrence rate after surgical treatment of DTF is closely related to margin status: R0 (negative margins) or R1 (microscopically positive margins). For small, well-defined DTFs without invasion of critical structures, complete tumor resection with R0 margins can be achieved. However, due to the locally aggressive behavior of DTF, large tumors with extensive invasion, particularly involving critical structures, are difficult and do not need to be completely resected, and maximal functional preservation should be prioritized. According to Bektas M (2023), the overall recurrence rate after surgery for DTF ranges from 15% to 40%, with higher rates observed in extra-abdominal DTFs (29.6% to 45.5%). Postoperative radiotherapy may reduce recurrence rates [5]. In 2017, Jassen ML published an analytical study on the role of margin status and radiotherapy in tumor recurrence after surgery in 1005 patients with primary DTF. The overall recurrence rate was 29%, and although it was lower in the group receiving combined radiotherapy, the difference was not statistically significant [11]. The study also concluded that the group with positive margins had a significantly higher recurrence rate compared to the group with negative margins. Another study in 114 patients reported similar findings; however, the authors recommended that achieving negative margins should not be pursued at all costs if it results in impairment of important functions. In some large, highly invasive tumors, positive margins may be accepted in order to preserve optimal function for the patient, particularly in tumors located in the head and neck region [12].

Another important issue in the management of DTF is the role of radiotherapy. Radiotherapy may be used as an adjuvant treatment in cases of incomplete resection or when the tumor is unresectable. However, the use of radiotherapy in young patients should be carefully considered due to the risk of long-term radiation-related complications [13]. These complications include soft tissue fibrosis, disturbances in bone and soft tissue development, as well as the risk of radiation-induced secondary malignancies. These risks are particularly concerning in young patients due to their longer life expectancy and the fact that the musculoskeletal system is still developing, resulting in a higher likelihood of cumulative radiation damage and late-onset complications. In our case, the patient was 19 years old, and the tumor was located in the cervical–supraclavicular region, where many important neural and vascular structures are concentrated. Radiotherapy in this region may increase the risk of soft-tissue fibrosis, nerve injury, and reduced shoulder–arm mobility, thereby leading to long-term impairment of upper limb function and quality of life. For these reasons, we decided not to recommend postoperative radiotherapy and instead selected an active surveillance strategy after surgery, with clinical and paraclinical follow-up.

4. CONCLUSION

Our case highlights several important points in the management of DTF. First, DTF in the cervical–supraclavicular region involving the brachial plexus is rare and poses significant surgical challenges. Second, maximal tumor resection while preserving important neural structures may result in favorable functional outcomes. Finally, in young patients, limiting the use of radiotherapy when possible may be a reasonable strategy to avoid long-term radiation-related complications, particularly when close postoperative follow-up can be ensured.

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