Desmoid-type fibromatosis of the head and neck region: A case report and brief review of the literature

WRYA N. SABIR[1], ABDULWAHID M. SALIH[2,3], ARI M. ABDULLAH[4,5], HIWA O. BABA[2], ABDULLAH A. QADIR[2], HARDI M. DHAHIR[2], REBAZ O. MOHAMMED[6], ASO S. MUHIALDEEN[2], SHKO H. HASSAN[2], KARZAN M. SALIH[2] and FAHMI H. KAKAMAD[3,7-9]

[1]Department of Pediatric Surgery, Smart Health Tower, Sulaymaniyah 46001, Iraq; [2]Department of Head and Neck Surgery, Smart Health Tower, Sulaymaniyah 46001, Iraq; [3]College of Medicine, University of Sulaimani, Sulaymaniyah 46001, Iraq; [4]Department of Histopathology, Smart Health Tower, Sulaymaniyah 46001, Iraq; [5]Department of Pathology, Sulaimani Teaching Hospital, Sulaymaniyah 46001, Iraq; [6]Department of Oncology, Smart Health Tower, Sulaymaniyah 46001, Iraq; [7]Department of Scientific Affairs, Smart Health Tower, Sulaymaniyah 46001, Iraq; [8]Department of Thoracic and Vascular Surgery, Smart Health Tower, Sulaymaniyah 46001, Iraq; [9]Kscien Organization for Scientific Research (Middle East Office), Sulaymaniyah 46001, Iraq

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Abstract. Desmoid-type fibromatosis (DTF) is a rare tumor characterized by locally infiltrating mesenchymal growth. Its occurrence in the head and neck region is rare. The present study describes the case of a male pediatric patient diagnosed with DTF involving the level II. A 2-year-old male patient presented with a progressively enlarging left submandibular mass over a period of 3 months. The results of hematological tests were normal. A neck ultrasound revealed a well-defined, lobulated hypoechoic left subcutaneous mass, measuring 38x31x27 mm, with mild vascularity and features suggestive of a suppurative or necrotic lymph node, accompanied by bilateral cervical lymphadenopathy with preserved morphology, with the largest node measuring 15x6 mm in the left level II. Computed tomography scan revealed no chest or abdominal abnormalities. The mass was completely excised, and a histopathological analysis confirmed DTF. The patient was discharged in good health. In addition, following a literature search, seven reported cases of DTF in the head and neck region were reviewed; the patients were aged 9 months to 42 years. DTF was found in the neck in 4 cases, in the mandible in 2 cases, and tongue in 1 case. Surgical excision led to uneventful outcomes for the majority of patients. DTF of the head and neck presents a complex clinical scenario. Its rarity and the challenges associated with diagnosis, location and treatment necessitate careful consideration, particularly in pediatric cases, in order to prevent unnecessary intervention and complications.

Correspondence to: Dr Fahmi H. Kakamad, College of Medicine, University of Sulaimani, HC8V+F66, Madam Mitterrand Street, Sulaymaniyah 46001, Iraq
E-mail: kakamad.fahmi@gmail.com

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Introduction

Desmoid-type fibromatosis (DTF) is a benign, locally infiltrating mesenchymal tumor that causes fibroblast and myofibroblast proliferation, and increased collagen production in deep musculoaponeurotic structures. It has intermediate biological characteristics between benign fibrous lesions and fibrosarcoma (1). The disease is extremely rare, accounting for only 0.03% of all neoplasms (2). Additionally, a notable predilection towards females has been observed (3). The etiology of DTF is multifaceted, possibly influenced by a combination of genetic predisposition, traumatic events and hormonal factors (4). DTF is classified into three subtypes based on their anatomical location: Abdominal wall, intra-abdominal, and extra-abdominal. Extra-abdominal desmoid tumors account for approximately one-third of all desmoid cases and most commonly arise in the shoulder, pelvic girdle and limbs. However, only 10-25% of cases occur in the head and neck region (5). Unlike other benign neoplasms in this area, desmoid tumors exhibit infiltrative growth and a marked propensity for local recurrence following surgery, underscoring the need for accurate diagnosis and appropriate management. Although complete resection with negative microscopic margins is considered the standard of care for DTF which is unresponsive to other treatments, achieving such margins in the head and neck is challenging due to the density of vital structures. This difficulty is even more pronounced in pediatric patients, where surgeons may hesitate to perform wide resections owing to the substantial risk of post-operative functional morbidity (3,4).

The present study describes the case of a male pediatric patient diagnosed with DTF involving the level II. The present case report was written in accordance with the CaReL guidelines, with filtering references to prevent citing contents in blacklisted journals (6,7).

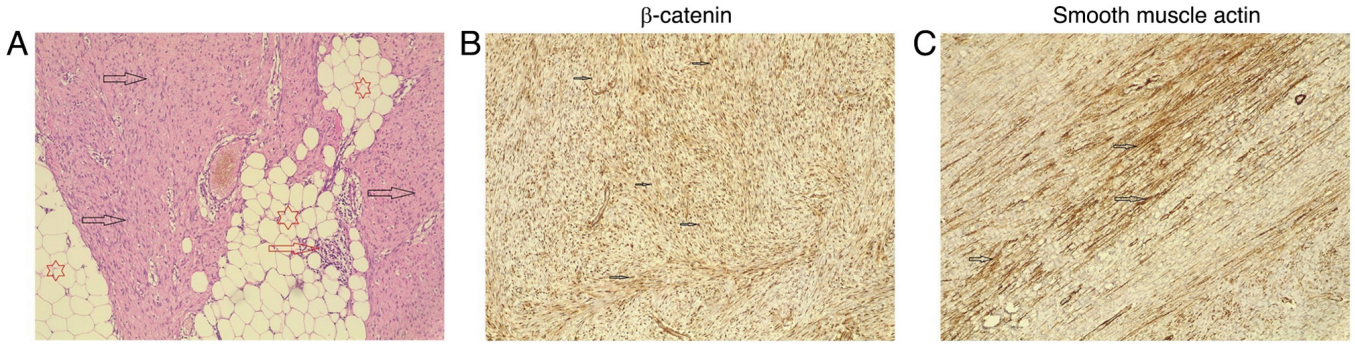

Figure 1. (A) Section illustrating a cellular area composed of bland-looking spindle cells (black arrows), infiltrating into adjacent lobules of adipocytes (red stars) with the presence of an aggregation of benign lymphoid cells (red arrow) (hematoxylin and eosin staining; magnification, x10). (B) β-catenin immune staining; black arrows indicate a nuclear staining pattern in spindle cells. (C) Smooth muscle actin; image indicates a cytoplasmic staining pattern in the spindle cells (black arrows).

Case report

Patient information. On December 20, 2023, a 2-year-old male patient was brought to Smart Health Tower, Sulaymaniyah, Iraq, presenting with a gradually progressing swelling in the left submandibular region that had developed insidiously over the previous 3 months.

Clinical findings. The neck mass was large, firm and non-tender, below the angle of the mandible on the left side of the neck.

Diagnostic assessment. The hematological test results of the patient were normal. A neck ultrasound revealed a well-defined, lobulated, predominantly solid hypoechoic left subcutaneous mass, measuring 38x31x27 mm, with mild vascularity, mild surrounding inhomogeneity and features suggestive of a suppurative or necrotic lymph node. There was bilateral cervical lymphadenopathy, with nodes of variable size, well-defined, with a cortical thickness <2 mm, and a preserved shape and hilar echotexture; the largest node measured 15x6 mm in the left level II. The thyroid, submandibular and parotid glands were normal, with no focal lesions (data not shown). A contrast-enhanced computed tomography (CT) scan of the chest and abdomen yielded normal findings (data not shown).

Therapeutic intervention. A surgical cervical exploration with lymph node biopsy was performed under general anesthesia with the patient placed in the supine position. A post-operative histopathological examination (HPE) was performed. The biopsy specimen was placed into tissue cassettes and processed using the DiaPath Donatello automated processor with a standard 11-h protocol involving graded alcohols, xylene and paraffin. Following paraffin embedding and trimming, blocks were sectioned at a thickness of 4-6 μm onto standard glass slides, incubated in an oven at 60˚C overnight, and stained with hematoxylin and eosin (H&E) on the DiaPath Giotto automated stainer using Gill II hematoxylin (1% for 10 min). The slides were then dried, coverslipped, and examined under a light microscope (Leica Microsystems GmbH). For immunohistochemistry, paraffin-embedded tissues were sectioned at 4-6 μm and mounted onto charged glass slides, followed by overnight incubation at 60˚C. Antigen retrieval was performed using the Dako PT Link system (Agilent Technologies, Inc.) by heating the sections to 100˚C for 5-10 min in either pH 6.0 or pH 9.0 retrieval solution, depending on the target antibody. The slides were then washed for 1 min in a 20 ml buffer solution (0.05 mol/l Tris/HCl, 0.15 mol/l NaCl, 0.05% Tween-20, pH 7.6) at room temperature. Hydrophobic wells were created using the Dako Pen (Agilent Technologies, Inc.). Endogenous peroxidase activity was blocked with 3% hydrogen peroxide. Primary antibodies [smooth muscle actin (mouse monoclonal, clone 1A4, 1:100, cat. no. M0851) and β-catenin (mouse monoclonal clone β-catenin-1, 1:100, cat. no. M3539) both from Dako; Agilent Technologies, Inc.] were applied at room temperature for 80 min, followed by incubation with a horseradish peroxidase-conjugated secondary antibody (1:200, cat. no. P0447, Dako; Agilent Technologies, Inc.) and diaminobenzidine (DAB) chromogen for 15 min each at room temperature. Counterstaining was performed with Gill II hematoxylin for 30 sec, after which the slides were dried and coverslipped. The post-operative HPE revealed a 4-cm, well-defined mass with a rubbery tan-white fibrotic texture. It was diagnosed as a low-grade spindle cell neoplasm, specifically a DTF, aided by immunohistochemical staining, which yielded positive results for smooth muscle actin and β-catenin (Fig. 1).

Follow-up and outcome. The post-operative period was uneventful, and the patient was discharged from the hospital in good health after 2 days. The follow-up treatment plan included regular clinical assessments and imaging as needed to monitor for recurrence, as part of standard care for DTF, which has a risk of local recurrence. Following a 6-month follow-up period, no recurrence was reported.

Discussion

Herein, a literature review was also conducted to identify reports on DTF of the head and neck through a Google Scholar search using the key word 'neck desmoid fibromatosis'. A total of 7 cases of DTF in the head and neck region were reviewed and these are summarized in Table I. Of the 7 cases, 4 patients were female and 3 patients were male, with ages ranging from 9 months to 42 years. The tumor in 4 cases was located in the neck, while the remaining tumors were

Table I. Summary of seven reported cases of desmoid fibromatosis of the head and neck region identified in the literature.

Authors	Age	Sex of included patient	Presenting compliant	Examination	Location	Imaging used for diagnosis (CT/MRI)	Management	Outcome	(Refs.)
Kant *et al*	22 years	Female	Intermittent neck pain, numbness, and weakness in her left shoulder	Weakness was evident in her left shoulder and left elbow, and sensory loss was noted in the left C3, C4, and C5 dermatomes	Neck	Well-defined lobulated multicompartmental mass along the left brachial plexus (C2-C6), occupying the paraspinal, prevertebral, and parapharyngeal spaces, displacing the trachea and esophagus rightward and the left carotid sheath anterolaterally.	Cervical exploration and subtotal decompression of the tumor were performed	Unknown	(4)
	36 years	Male	Gradually progressing swelling on the left side of his neck	Neck mass was large, firm, and non-tender, extending from the angle of the mandible to the clavicle on the left side of the neck	Neck	Multilobulated isointense mass on the left neck extending along the left brachial plexus, medially into the neural foramina, and inferiorly into the mediastinum.	Unknown	Unknown	(4)
Miyashita *et al*	9 months	Female	A tumor rapidly enlarged, with part protruding from the tongue	The patient was unable to fully close her mouth, and swallowing was aided by compensatory motion of the unaffected side	Tongue	Whole-body CT examination revealed no signs of any other tumoral lesions. MRI revealed a large mass with contrast enhancement on the right side of the tongue. MRI of the sagittal plane did not show the possibility of infiltration into the root of the tongue.	A partial glossectomy with a 5-mm safety margin and simultaneous reconstruction with a local flap were performed	Good outcome without recurrence after 1 year	(8)
Bouatay *et al*	38 years	Female	Recurrent desmoid fibromatosis after 1 month of management	Painless, hard, non-tender right laterocervical swelling (8 cm), fixed to the skin and deep structures	Neck	Cervical CT scan revealed a soft tissue mass infiltrating the deep lateral cervical muscles and surrounding superficial and deep fat.	Complete tumor resection	No local recurrence was observed after 15 months	(11)

Table I. Continued.

Authors	Age	Sex of included patient	Presenting compliant	Examination	Location	Imaging used for diagnosis (CT/MRI)	Management	Outcome	(Refs.)
Alherabi *et al*	42 years	Female	Neck mass gradually increasing in size	Hard, non-tender swelling in the left posterolateral upper neck, fixed to the skin and deep structures	Neck	Neck CT scan revealed a soft tissue mass in the upper neck, lateral to the sternocleidomastoid muscle, infiltrating deep lateral cervical muscles and adjacent superficial and deep fat.	Mass widely excised with a small remnant at the carotid bifurcation; 16-month CT showed slow regrowth, treated with 60 Gy radiotherapy. The case encountered temporary partial facial nerve paralysis	Good after a 2-year follow-up with significant regression of the tumor	(14)
Sato *et al*	3 years	Male	Swelling of submandibular region for 1 month	Inspection and palpation revealed a 50x35 mm bony-hard, poorly mobile mass arising from the left mandibular angle, expanding anteroinferiorly	Mandible	MRI revealed a 50x45x32 mm left mandibular tumor with unclear margins and high T2 signal; CT scan revealed moth-eaten mandibular destruction.	Surgical excision was performed	Good without recurrence after 25 months	(5)
Said-Al-Naief *et al*	8 years	Male	A rapidly enlarging, painless mass along the right mandibular border for two months.	Slight expansion of the right mandibular border with overlying soft tissue fullness.	Mandible	CT scan revealed an 8 mm right mandibular body mass with outer cortical destruction.	Mass resection with wide margins of the right inferior mandibular border was performed.	Good without recurrence after 4.5 years	(15)

CT, computed tomography; MRI, magnetic resonance imaging; HPE, histopathological examination.

found in the mandible (n=2) and tongue (n=1). Among the cases reviewed herein, the neck tumors generally progressed gradually, occasionally causing symptoms, such as numbness, pain and weakness, whereas tumors in other locations exhibited a more rapid progression. The preferred imaging modalities for these tumors were a CT scan and magnetic resonance imaging (MRI). Surgical exploration with mass excision and clear safety margins was the standard approach for managing these tumors. The majority of patients experienced uneventful outcomes following surgery, apart from 1 patient who developed temporary partial facial nerve paralysis.

DTF of the head and neck is a rare benign mesenchymal tumor with an annual incidence of 2-4 cases per million individuals (2,8). In the case series study by Hoos *et al* (9), a sex predilection for females was noted, whereas in the literature review performed in the study by Miyashita *et al* (8), including 141 pediatric patients with DTF of the head and neck aged birth to 18 years, no sex predominance was observed. In the literature review performed in the study by Miyashita *et al* (8), the mandible was the most common site of involvement, accounting for 25% of cases. Additional reported sites included the submandibular region, the infratemporal fossa, the neck, the peritracheal area and the paraspinal region (8).

Empirical evidence indicates that among DTFs involving the neck, the anterolateral aspect is a common site, with the majority of patients presenting with a painless mass (60-94%). However, neurogenic symptoms, such as pain or focal motor deficit may be observed in a smaller proportion of cases (16-36%) (10). Compression of the brachial plexus can result in symptoms, such as tingling, paresthesia, numbness and weakness in hand muscles (1). A rapidly enlarging DTF was also reported in a pediatric case in the study by Miyashita *et al* (8). The case in the present study was a 2-year-old male presenting with a painless, slow-growing swelling in the left submandibular region, which progressed over a period of 3 months.

The diagnosis of DTF in the head and neck region is primarily based on a clinical presentation, imaging studies and biopsy. While radiographs may be a useful initial diagnostic tool, their findings can vary, and a definitive diagnosis usually requires HPE (10,11). The accurate assessment of the extent of the tumor and its relation to surrounding structures is crucial for effective management. Advancing imaging techniques, such as neck CT scans and MRI, play a key role in such an assessment. MRI, in particular, is preferred for its superior soft tissue contrast, which aids in more precise tumor delineation and reduces the risk of damaging critical structures during surgery (11). Typically, DTF exhibits intermediate signal intensity on T1-weighted images (T1WIs), similar to muscle tissue, and on T2-weighted images (T2WIs), where the signal is lower than that of fat, but higher than that of adjacent muscle. Fat-suppressed T2-weighted images often exhibit increased signal intensity within these lesions. Areas with a low signal intensity within DTF correspond to regions of higher collagen content. Post-contrast imaging usually reveals moderate to marked heterogeneous enhancement, reflecting the vascularity of the lesion. In the case in the present study, a neck ultrasound revealed a well-defined, lobulated, predominantly solid hypoechoic left subcutaneous mass, measuring 38x31x27 mm, with mild vascularity. Chest and abdominal contrasted CT scans yielded normal findings.

The primary clinical differential diagnoses include soft-tissue sarcoma, lymphoma, myositis ossificans and arteriovenous malformation. Imaging often rules out the latter conditions; however, distinguishing desmoid tumors from soft tissue sarcomas can be challenging (12). Desmoid tumors exhibit a spectrum of clinical behaviors, ranging from indolent growth to aggressive local infiltration, which significantly influences management strategies. Aggressive variants, characterized by rapid growth, local invasion and a higher risk of recurrence, often require multimodal treatment. Post-operative radiation therapy (RT) is particularly considered in cases where achieving negative surgical margins is challenging or where tumors are located near critical structures, making complete excision infeasible without undue morbidity (1,13). Despite this, the role of RT remains controversial due to the benign histological nature of desmoid tumors and the potential for significant complications, coupled with a lack of prospective clinical trials assessing its efficacy in reducing recurrence rates (11,14,15). Current evidence is derived from retrospective studies, which suggest that combining RT with surgery may improve local control rates, particularly in cases where wide margins are difficult to achieve (8). Surgical resection has traditionally been the primary treatment for primary and recurrent desmoid tumors, aiming for tumor-free margins. A 5-year local control rate of 80% with negative-margin resection has been reported. However, some experts argued that achieving negative margins may lead to unnecessary complications and may not prevent local recurrence (11). The recurrence rate among pediatric patients undergoing surgery has been reported as 27.2% (8).

For benign or less aggressive variants, which demonstrate slow growth and minimal recurrence risk, a more conservative approach, such as watchful waiting, is gaining acceptance, particularly for asymptomatic cases or tumors in anatomically challenging locations. Given the challenges of balancing treatment efficacy with minimizing overtreatment or unnecessary morbidity, the management of desmoid tumors underscores the need for a multidisciplinary approach. In pediatric patients with DF that is unresponsive to non-surgical therapies, surgeons may hesitate to pursue wide resections due to the substantial risk of postoperative morbidity. When a large tumor is located near vital structures, achieving adequate surgical margins can be particularly challenging (8). The literature review performed in the study by Miyashita *et al* (8) revealed post-operative complications in 13 patients (10.4%). Trismus was the most frequent (n=6). A total of 2 patients developed secondary papillary carcinoma following radiation therapy. Additional complications included osteomyelitis (n=2), mild ptosis resulting from facial nerve sectioning (n=1), restricted neck mobility (n=1) and Claude-Bernard-Horner syndrome (n=1) (8). Individualized treatment planning, guided by the biological behavior of the tumor, is essential to optimize patient outcomes while addressing the limitations of current evidence and therapeutic options (10,15). In the present case report, the mass was successfully excised under general anesthesia with no complications. No recurrence was reported following 6 months of follow-up. The limitations of the present case report include the inability to retrieve the ultrasound figures due to poor archiving and CT scan figures for the case, as they were conducted at an external facility.

In conclusion, DTF of the head and neck presents a complex clinical scenario. Its rarity and the challenges associated with diagnosis, location, and treatment necessitate careful consideration, particularly in pediatric cases, in order to prevent unnecessary interventions and complications. Ongoing research is thus warranted to define standardized treatment protocols and improve therapeutic outcomes for this challenging condition.

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Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

FHK and AMS were major contributors to the conception of the study, as well as to the literature search for related studies. WNS, AAQ and SHH contributed to the clinical management of the patient, assisted in data acquisition and interpretation, and participated in the literature review and manuscript preparation. HOB, HMD, ROM, ASM and KMS contributed to the conception and design of the study, the literature review, the critical revision of the manuscript, and in the processing of the table. SHT was the radiologist who performed the assessment of the case. AMA was the pathologist who performed the diagnosis of the case. FHK and AMS confirm the authenticity of all the raw data. All authors have read and approved the final manuscript.

Ethics approval and consent to participate

Written informed consent was obtained from the patient's parents for participation in the present study.

Patient consent for publication

Written informed consent was obtained from the patient's parents for the publication of the present and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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