Desmoid Tumor Formation following Posterior Spinal Instrumentation Placement

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Abstract

Study Design Case report.

Objective The objective of the article is to illustrate a case of desmoid tumor (DT) formation after posterior instrumentation of the thoracic spine.

Methods A 57-year-old woman presented with lower extremity clumsiness, balance, and ambulation difficulty resulting from spinal cord compression due to an upper thoracic atypical vertebral hemangioma. Ten months after undergoing embolization, resection, and placement of instrumentation for this lesion, the patient developed a growing mass at the rostral end of the incision. Biopsy revealed desmoid fibromatosis. The mass was removed via an en bloc resection. Histology revealed an infiltrative DT above the laminectomy site abutting the instrumentation.

Results At 2-year follow-up, there was no evidence of recurrence of the tumor.

Conclusion Paraspinal DTs have been reported in the literature to develop after surgical procedures of the spine. Often times, patients attribute swelling or fullness at the site of their surgery to scar tissue formation or instrumentation. One must consider the possibility of a DT in the setting of reported surgical site fullness or mass after spine surgery. It is thought that postoperative inflammation present in the surgical bed may promote formation of DTs. Instrumentation may also contribute to inflammation and increase the likelihood of developing a DT. Generous margins must be taken to prevent recurrence.

Keywords
- desmoid tumor
- corpectomy
- aggressive fibromatosis

Introduction

Desmoid tumors (DTs), also known as aggressive fibromatoses, are characterized as rare, slow-growing tumors. Although strictly benign, they are locally invasive with a high rate of recurrence even after seemingly total resection.[1] DTs typically develop sporadically, often in the abdomen, hip/buttocks area, and shoulder girdle. Some populations are at higher risk, including individuals with episodes of antecedent trauma at the site of the tumor.[2] A particularly rare subset of these cases includes DTs that develop adjacent to the spinal column. A total of five reported cases have described this phenomenon in the absence of Gardner syndrome, a variant of familial adenomatous polyposis (FAP), or any other predisposing genetic diseases.[3–7]

This report describes an instance of postoperative aggressive fibromatosis that developed 10 months after corpectomy and spinal fusion for treatment of an atypical vertebral hemangioma in the thoracic spine.

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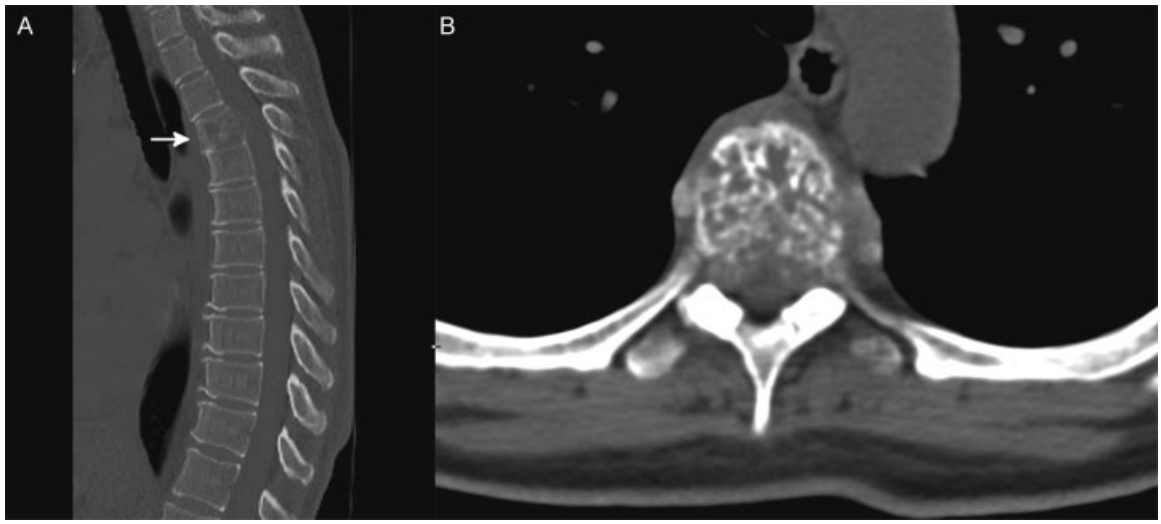

Fig. 1 Preoperative images depicting the original hemangioma. (A) Sagittal computed tomography (CT) demonstrating the hemangioma in the T4 vertebral body (arrow). (B) Axial CT at the T4 vertebral level.

Case Report

A 57-year-old woman presented with lower extremity clumsiness, balance, and ambulation difficulty resulting from spinal cord compression due to an upper thoracic atypical vertebral hemangioma. The patient underwent embolization followed by resection (►**Fig. 1**). The resection involved a transpedicular approach for a T4 corpectomy, placement of an expandable cage extending from T3 to T5 and instrumented posterior fixation from T2 to T7. The patient made a good recovery from the uncomplicated procedure.

Ten months postoperatively, the patient reported moderate pain at the rostral end of the surgical incision. A mass reported by the patient to have enlarged was found on palpation, just superior to the surgical incision. As the mass continued to grow, the patient suffered from increasing local pain and pressure, but there were no neurological deficits.

Magnetic resonance imaging (MRI) revealed a $5.7 \times 1.6 \times 3.1$ cm T2 hyperintense mass within the right subcutaneous tissues and paraspinal muscles at the T1–T3 levels (►**Fig. 2**). The mass was located superficial to a pedicle screw at the rostral end of the fusion construct. The tumor was removed via an en bloc resection. Negative margins were achieved through added removal of the spinous processes of T1–T2 and part of C7. The fully excised tumor was measured to be 4.5 cm in diameter.

The firm rubbery mass was in some areas histologically compact and appeared to exclude preexisting elements. Elsewhere was infiltrative and trapped skeletal muscle fibers and adipose tissue. The tumor was composed largely of dense collagen and fibroblasts. The degrees of cellularity and cytological atypia varied from region to region, but both were low in most areas. In a few regions, however, there was a modest degree of cytological atypia and low-level mitotic activity. Mitoses were absent in paucicellular, more fibrotic areas (►**Fig. 3**). Immunostaining for β-catenin was confined to the cytoplasm in most cells, but there were scattered cells in which nuclei were also positive.

Serial MRI was performed to follow for tumor recurrence. At 2-year follow-up, there has been no evidence of recurrent disease. The patient has done well and is back to work as a nurse educator.

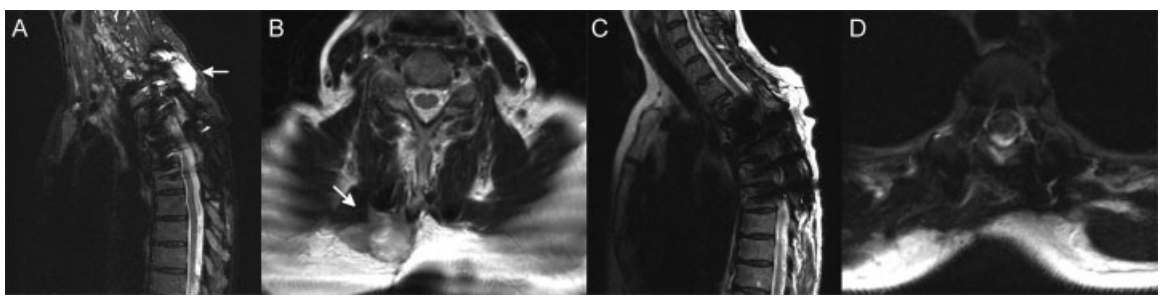

Fig. 2 Magnetic resonance images (MRI) showing the postlaminectomy desmoid tumor (DT) and results after surgical excision. (A) Preoperative sagittal STIR MRI showing the DT (arrow). (B) Preoperative axial T2-weighted MRI of the same tumor (arrow). (C) Postoperative sagittal T2-weighted MRI demonstrating absence of tumor. (D) Postoperative axial T2-weighted MRI demonstrating absence of tumor.

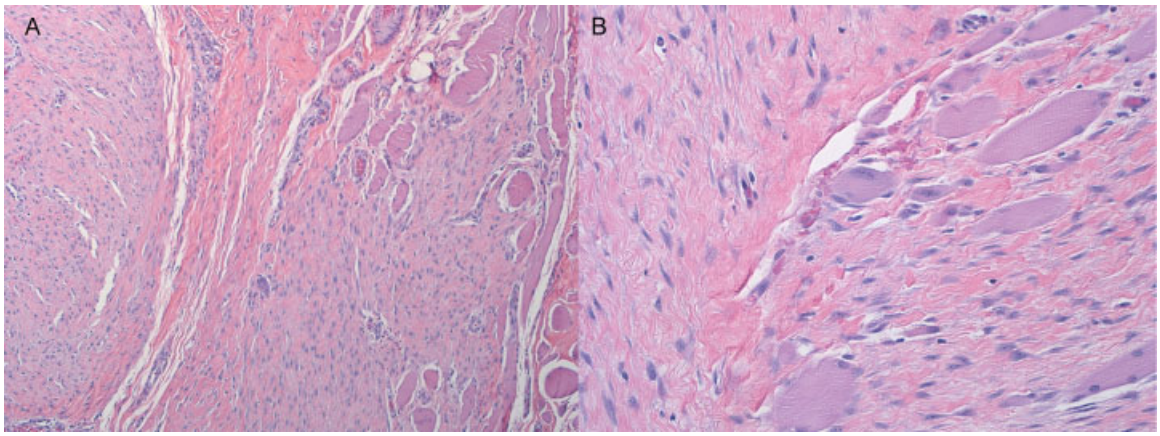

Fig. 3 Hematoxylin and eosin stain of the desmoid tumor. (A) The paucicellular, somewhat fascicular and lobular mass incorporates regional tissues such as skeletal myocytes (40×). (B) Cell density here is low and there is only slight-to-moderate cytological atypia. Other, more cytologically atypical, areas were somewhat more cellular. Myocytes are trapped in the infiltrating lesion (100×).

Discussion

Aggressive fibromatosis comprises 0.03% of all neoplasms and less than 3% of all soft tissue tumors.[2] Several conditions such as FAP and other hereditary cancer syndromes predispose individuals to developing these fibroblastic growths.[8] A summary of the five previously reported cases of DT formation soon after laminectomy in patients without a unique predisposing condition are presented in ►**Table 1**.[3–7]

When comparing these cases, it is clear that there are striking similarities. All cases occurred in adult women ranging in age from 39 to 50 years, with our patient, a 57-year-old woman, having similar demographics. This is concordant with previous studies that have asserted that 67 to 80% of all DTs arise in women, with 50% between the ages of 30 to 50 years. It has been previously suggested that aggressive fibromatosis may be hormonally related.[2] In fact, middle-aged mothers may be at heightened risk because of characteristic hormone profiles.

In addition, none of the five previous cases developed neurological deficits as a result of the DT. Two patients did have mild residual neurological side effects as a result of the initial laminectomy procedure, but these symptoms were unrelated to the DT. Our patient was similarly neurologically asymptomatic.

The number of reported cases of paraspinous aggressive fibromatosis cases occurring postlaminectomy indicates that this may not be as rare of a phenomenon as originally suspected. The scar tissue surrounding a laminectomy may be predisposed to neoplastic transformation due to underlying inflammation from both the treated malignancy and tissue manipulation from surgery.

Studies have elucidated some of the mutations that lead to sporadic DTs. Recent studies have demonstrated that sporadic DTs often have mutations in adenomatosis polyposis coli and β-catenin genes. A study by Lazar et al demonstrated that 85% of sporadic DTs have mutations in the *CTNNB1* gene, which encodes β-catenin. Furthermore, different *CTNNB1* mutations were associated with differing rates of progression and recurrence.[9]

β-catenin has been observed to play a significant role in postfracture bone healing. A study by Chen et al demonstrated that β-catenin signaling is activated during bone healing. Furthermore, resultant T cell factor-dependent transcription is only activated in later stages of repair, after mesenchymal cells have differentiated into fibroblastic cells.[10] This would suggest a unique molecular and cellular environment within the surgical bed during both the acute and chronic stages of healing. Inflammatory cytokines unregulated at both these time points likely can potentiate DT growth. Theoretically, any bone fracture has the potential to trigger DT growth that could explain the development of aggressive fibromatosis after clavicle and radius fractures.

DTs are often difficult to treat because of their proliferative nature and capacity for local invasion. Treatment becomes even more challenging when these masses are found proximal to sensitive anatomical structures, such as in the spine. In the presented case and five others, gross total resection with negative margins was elected. This was seen to be the best option given the high local recurrence rate associated with positive margin resections (54% over 10 years) as compared with those with negative margins (27% over 10 years). If the mass were located proximal to functionally important anatomic entities such that negative margins were impossible to achieve, adjuvant radiation therapy (25% recurrence over 10 years) would have been another option.[1]

Conclusion

Middle-aged women undergoing laminectomies have been observed to develop postoperative DTs of the spine. Although these tumors do not cause neurological deficits, they tend to be locally invasive and are best treated with wide excision with negative margins.

Although it has been speculated that hormonal triggers could be involved in postsurgical DT formation, it is also likely that inflammatory cytokines released in the surgical bed during the acute and chronic phases of healing also play a role in growth of aberrant tissue. This may be particularly true

Table 1 Summary of present previously reported postoperative spinal desmoid tumors

Author	Age/Sex	Location of laminectomy	Original condition	Instrumentation	Neurological deficit	Location of tumor	Treatment
Gonatas 1961[3]	45 F	Cervical	Intervertebral disc	None	None	Adherent to rhomboid and levator scapulae and scapula	Excised in entirety
Wyler and Harris 1973[7]	39 F	C6-T1 hemilam. C8-T2 dorsal rhizotomy	Dysesthesia in left elbow with radiation to forearm	No	None	Paraspinous muscles	En bloc after recurrence
Lynch et al 1999[5]	49 F	T9-T11	Thoracic meningioma	None	None[a]	Paraspinous	Surgical resection with wide margins
Güzey 2006[4]	50 F	L4-L5	Spondylolisthesis	Yes	None	T12, L1 spinous processes, Rostral margin	Total resection with 1 cm safety margin
Sevak et al 2012[6]	48 F	C6-T1	Extradural schwannoma	Yes	None[a]	Subcutaneous	Wide local excision with right trap; myocutaneous flap reconstruct
Current	57 F	T4	Vertebral hemangioma	Yes	None	Subcutaneous Rostral Margin	En bloc resection

[a]Some mild postoperative (procedure before development of desmoid tumor) paresthesias or weakness.
The studies are listed chronologically.

with the continued presence of surgical instrumentation for fixation, which represents foreign material within the surgical bed.

Disclosures
Dr. Witham receives research support from Eli Lilly and Company for spinal fusion research, but this relationship is not directly relevant to the manuscript that is being submitted. This clinical case report and manuscript has been drafted with strict adherence to the highest research ethics and integrity standards.

References

1 Ballo MT, Zagars GK, Pollack A, Pisters PWT, Pollack RA. Desmoid tumor: prognostic factors and outcome after surgery, radiation therapy, or combined surgery and radiation therapy. J Clin Oncol 1999;17(1):158–167
2 Reitamo JJ, Häyry P, Nykyri E, Saxén E. The desmoid tumor. I. Incidence, sex-, age- and anatomical distribution in the Finnish population. Am J Clin Pathol 1982;77(6):665–673
3 Gonatas NK. Extra-abdominal desmoid tumors. Report of six cases. Arch Pathol 1961;71:214–221
4 Güzey FK, Emel E, Bas NS, Ozkan N, Turgut H, Sel B. Aggressive postoperative lumbar fibromatosis after the placement of instrumentation for treatment of spondylolisthesis. Case report. J Neurosurg Spine 2006;4(4):338–341
5 Lynch JJ, Parvizi J, Scheithauer BW, Krauss WE. Development of postoperative fibromatosis after resection of an intraspinal meningioma. Case report. J Neurosurg 1999;90(1, Suppl):121–124
6 Sevak S, Blount AL, Cottingham S, et al. Fibromatosis of the cervical region following laminectomy: a case report and literature review. Spine 2012;37(7):E456–E459
7 Wyler AR, Harris AB. Recurrent desmoid tumor following cervical laminectomy. Case report. J Neurosurg 1973;39(1):114–116
8 Gurbuz AK, Giardiello FM, Petersen GM, et al. Desmoid tumours in familial adenomatous polyposis. Gut 1994;35(3):377–381
9 Lazar AJ, Tuvin D, Hajibashi S, et al. Specific mutations in the beta-catenin gene (CTNNB1) correlate with local recurrence in sporadic desmoid tumors. Am J Pathol 2008;173(5):1518–1527
10 Chen Y, Whetstone HC, Lin AC, et al. Beta-catenin signaling plays a disparate role in different phases of fracture repair: implications for therapy to improve bone healing. PLoS Med 2007;4(7):e249

Commentary

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Postsurgical desmoid formation at the site of surgery is not a common pathology. Puvanesarajah et al describe a case of a 57-year-old woman who underwent resection and stabilization because of spinal cord compression due to a high thoracic (T4) hemangioma. However, the patient's neurologic symptoms improved. Ten months after the surgery, they observed a rapidly growing mass in the surgical scar, which was histologically determined to be a desmoid fibromatosis. They illustrate nicely the preoperative pathology and the desmoid tumor (DT) in the rostral part of the pathology. Performing en bloc resection of the DT with strict local control of the disease, succeeded in preventing local recurrence after 2 years.

DTs are benign, deep-seated monoclonal myofibroblastic neoplasms that grow slowly, infiltrative and arise from musculoaponeurotic stromal elements. In the past few years, the term "aggressive fibromatosis" also has come into use. The etiology of these tumors is likely multifactorial. Genetic, endocrine, and physical factors play a role because abdominal wall tumors often arise in young parous women following childbirth or in a postsurgical scar. DTs also occur commonly as part of hereditary syndromes, such as familial adenomatous polyposis (FAP), which are often associated with a germline mutation in the adenomatosis polyposis coli (APC) gene.[1]

As the authors pointed out, there are just five reported cases in the literature that describe de novo DT formation in the postsurgical scar in patients with an absence of any predisposing genetic syndromes.

There are some generally accepted principles regarding the treatment of DTs that are as follows.[2]

1. When medically and technically feasible, growing and symptomatic DTs should be treated surgically.[3]
2. En bloc resection of these tumors give the lowest rate of local recurrence, while positive surgical margins or intralesional resections show a much higher rate of recurrence.[4]
3. In the case of oncologically improper surgical resection, adjuvant radiotherapy would be an option.[5]

The same principles are dominant in postsurgical desmoid-formation. This special appearance in the postoperative scar was described after breast,[6,7] abdominal,[8] and other orthopedic[3] procedures. According to the literature, the first detection of the tumor formation showed a wide range, between 10 months and 7 years after the local surgery. The first clinical sign is a growing, well palpable (most frequently painless) tissue in the postoperative scar or inside the muscles dissected during the surgical approach. The myofibroblast is the cell considered to be responsible for the development of tumorous malformation, but the etiological factor to initiate this process is unknown (As the authors indicated, probably the activation of β-catenin signaling by different local tissue-healing factors could result in aggressive fibroblast proliferation, supported or promoted by inflammatory cytokines in the surgical bed).

The authors listed five previously reported postoperative spinal DTs. All of them were treated surgically by en bloc resection without local recurrence. In certain cases, after the extensive resection, a myocutan flap was necessary for the local soft tissue reconstruction.

In the cases described in the literature, the local pain caused by the postsurgical desmoid formation is more significant in the breast and the abdominal wall even in the earlier stage with smaller tumor size, whereas after spine surgery, the level of local pain caused by the tumors with paraspinal locations shows a very wide range. (In one case in our institutional experience, a 47-year-old female patient returned 11 months after a lower lumbar instrumented posterolateral fusion with a large, well palpable mass—later histologically proved desmoid—which was painless at the time of the recognition.)

The functional disturbances due to the tumor growth highly depend on the region the spinal surgery was performed, the muscle, and where the tumor appears first. Restricted spinal range of motion (especially with increasing local pain provoked by the movement) appearing after a certain stage of normal functional rehabilitation could also indicate abnormal tissue changes in the mass of the postoperative scar or the surrounding paraspinal muscles. In the cervical spine (despite the benign histological appearance and negligible metastatic potential), the growing DT infiltrating the surrounding tissues could lead to deformity, morbidity, and mortality resulting from pressure effects and also by the potential obstruction of vital structures and organs.[9]

The authors recognized and diagnosed the pathology in proper time and selected the state-of-the-art treatment method. The successful postoperative course and the lack of the local recurrence after 2 years show effective decision making.

References

1 Escobar C, Munker R, Thomas JO, Li BD, Burton GV. Update on desmoid tumors. Ann Oncol 2012;23(3):562–569
2 Berri RN, Baumann DP, Madewell JE, Lazar A, Pollock RE. Desmoid tumor: current multidisciplinary approaches. Ann Plast Surg 2011;67(5):551–564
3 Gebhart M, Fourmarier M, Heymans O, Alexiou J, Yengue P, De Saint-Aubain N. Development of a desmoid tumor at the site of a total hip replacement. Acta Orthop Belg 1999;65(2): 230–234
4 Dalén BP, Bergh PM, Gunterberg BU. Desmoid tumors: a clinical review of 30 patients with more than 20 years' follow-up. Acta Orthop Scand 2003;74(4):455–459
5 National Comprehensive Cancer Network (NCCN). Guidelines for Soft Tissue Sarcomas, Version 1. 2011. Available at: www.nccn.org. Accessed March 13, 2011
6 Mátrai Z, Tóth L, Gulyás G, Szabó E, Szentirmay Z, Kásler M. A desmoid tumor associated with a ruptured silicone breast implant. Plast Reconstr Surg 2011;127(1):1e–4e
7 Chummun S, McLean NR, Abraham S, Youseff M. Desmoid tumour of the breast. J Plast Reconstr Aesthet Surg 2010; 63(2):339–345
8 Wanjeri JK, Opeya CJ. A massive abdominal wall desmoid tumor occurring in a laparotomy scar: a case report. World J Surg Oncol 2011;9:35
9 Lewin JS, Lavertu P. Aggressive fibromatosis of the prevertebral and retropharyngeal spaces: MR and CT characteristics. AJNR Am J Neuroradiol 1995;16(4, Suppl):897–900

Editorial Perspective

This case report of aggressive fibromatosis in the posterior cervico-thoracic junction following an otherwise uneventful hemangioma resection underscores a number of important considerations:

1. It is helpful to have advanced postoperative imaging early after tumor resections of the spine as a baseline comparison study for any recurrences, reconstructive failures, or as happened in this case, a secondary neoplasia. A CT scan is most commonly chosen for this type of reference study (which is also helpful in checking adequacy of resection margins among other factors), but an MRI scan may be helpful by about 3 months after surgery when soft tissue lesions are of concern and the initial postsurgical changes have largely settled down.
2. This case also underscores the value of scheduled longer term follow-up following neoplasia care of the spinal column, as pointed out in the commentary by Dr. Varga.
3. There is hope that members of the AOSpine International community who increasingly treat spinal neoplasia join the AOSpine Neoplasia Knowledge Forum to place not only routine cases into their well-governed registry but especially rare cases like this one, preferably with tissue samples to allow for genetic studies later on. This may offer improved insights into the oncogenic circumstances of certain forms of neoplasia, such as the one found described here.

EBSJ thanks the authors for their contribution and Dr. Varga for his thoughtful commentary.