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Case Report

Desmoid fibromatosis following surgical resection of spinal meningioma ☆

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ABSTRACT

A 42-year-old female patient with a history of cervicothoracic junction meningioma World Health Organization grade I, resected in early 2011, was admitted to the hospital with intractable headache and lower extremity weakness. Magnetic resonance imaging (MRI) showed an epidural mass compressing the spinal cord at the prior surgical site, which was interpreted as recurrent meningioma. Following surgical resection, histopathological analysis revealed desmoid fibromatosis (desmoid tumor). In retrospect, dynamic contrast-enhanced magnetic resonance imaging performed prior to surgery should have allowed for prospective exclusion of meningioma as the recurrent mass and suggested an alternative diagnosis such as post-traumatic desmoid fibromatosis or the need for biopsy to confirm diagnosis.

Introduction

MacFarlane first described a desmoid tumor in 1832 [1] and 6 years later Müller coined the term desmoid, meaning "tendon-like" [2,3]. Desmoid tumors make up 0.03% of neoplasms, and only 2-4 cases per million people are reported each year [4]. Desmoid fibromatosis (aggressive fibromatosis) is a type of benign mesenchymal neoplasm that arises from the fibroblasts and myofibroblasts in fascia, aponeuroses, and muscle connective tissue as a clonal proliferation of spindle cells [3,5,6]. Desmoid tumors are classified based on their location, with subcategories of abdominal, extra-abdominal, and intra-abdominal [7]. The majority of these tumors are seen in patients within the age range of 15-60 years old, with the peak incidence in females between the ages of 25-35 years [2,8].

Desmoid tumors are associated with familial adenomatous polyposis (FAP) in the genetic disorder Gardner Syndrome, yet can also arise sporadically [9]. Trauma, hormonal responses, and a genetic predisposition are thought to play a role in the development of desmoid tumors. However, the exact etiology remains poorly understood [10,11]. Although lacking metastatic potential, desmoid tumors invade locally and have high recurrence rates linked with microscopic surgical

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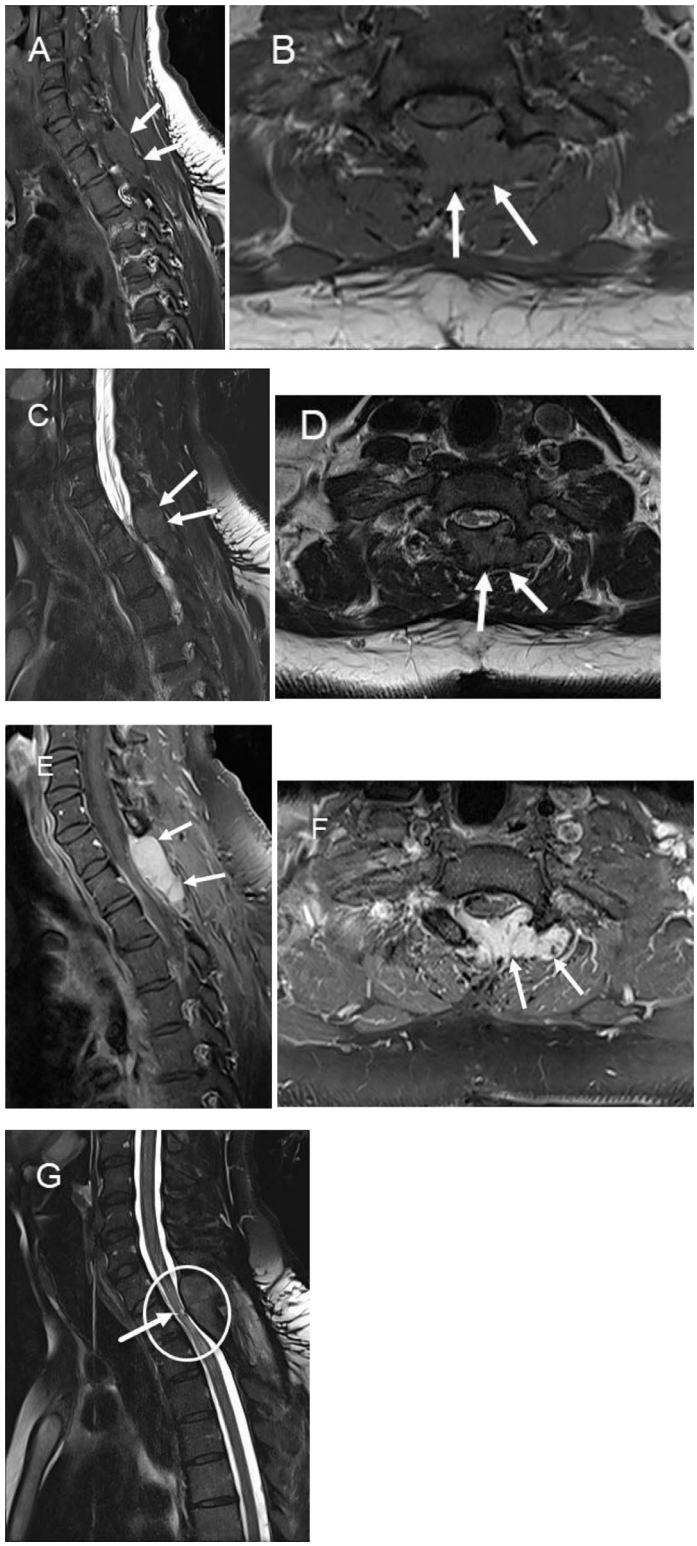

Fig. 1. – T1-weighted MRI of the spine in the sagittal (Fig. 1A) and axial (Fig. 1B) planes show a well-defined mass *(arrows)* along the dorsal aspect of the upper thoracic cord extending from C7-T1 to the inferior portion of T2 with infiltration of the adjacent left paraspinal musculature. The mass demonstrates signal intensity similar to that of the surrounding muscles. The mass appears isointense/slightly hyperintense to the surrounding muscles on T2-weighted MRI sequences (Fig. 1C & D). Following intravenous contrast administration, the mass expresses heterogeneous contrast enhancement (Fig. 1E & F). Sagittal T2-weighted MRI of the spine (Fig. 1G) shows slight compression and anterior displacement of the adjacent thoracic spinal cord *(circle)*. The compressed cord demonstrates slightly increased signal intensity *(arrow)*.

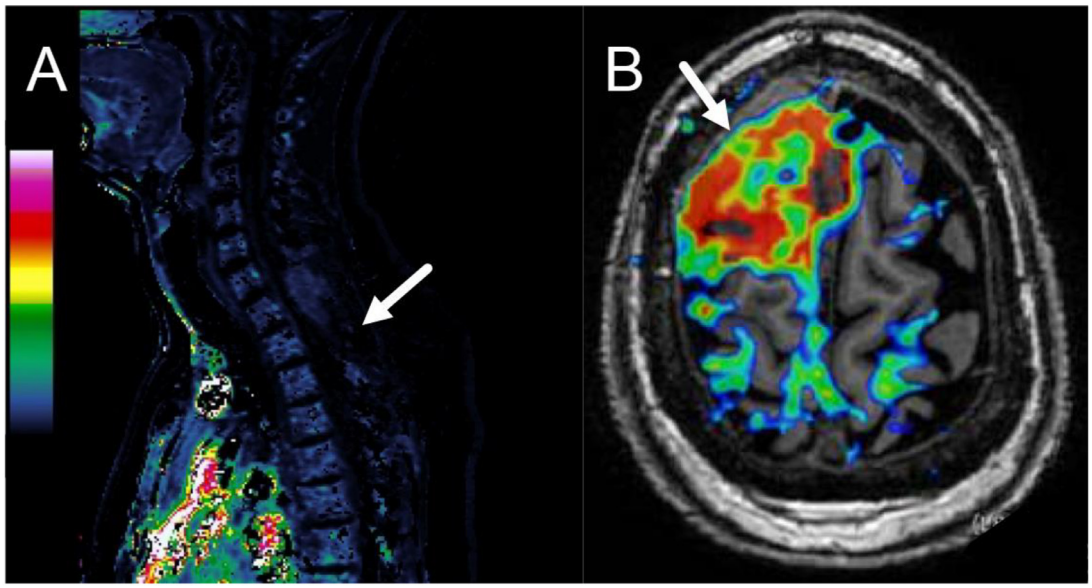

Fig. 2. – Sagittal DCE-MRI projection of the thoracic spine (Fig. 2A) demonstrates a low K^{trans} enhancing mass (K^{trans} = 0.0289 $min^{-}1$) along the dorsal aspect of thoracic cord extending from T1-T2 level *(arrow)*. While axial DCE-MRI projection of the brain (Fig. 2B) in an unrelated patient shows a predominantly right-sided extra-axial high K^{trans} enhancing mass correlating with a highly vascularized neoplasm found later to be WHO II meningioma *(arrow)*.

margins status. Leithner et al reported a 27% recurrence rate with wide or radical microscopic surgical margins and a 72% recurrence rate with a marginal or intralesional excision [12].

We present the case of a 42-year-old female with a history of WHO 1 intradural spinal meningioma at the level of T1-T2 who developed desmoid fibromatosis in the resection bed 6 years after surgery. Initially, the enhancing mass was assumed a recurrent spinal meningioma. However, histopathological analysis of the surgical specimen revealed a desmoid tumor presumed to be caused by iatrogenic trauma.

Case report

A previously healthy 42-year-old female with a history of WHO 1 spinal meningioma at the level of the T1-T2 vertebrae presented with intractable occipital headaches and lower extremity weakness. She was admitted to our institution to receive a second opinion at the suggestion of her local healthcare provider after MRI revealed a dorsal dural-based lesion at T1-T2 causing spinal cord compression concerning for recurrence at the surgical site..

Imaging

MRI of the cervical and thoracic spine showed a well-defined mass with signal intensity on T1-weighted (T1W) MRI sequences similar to that of the surrounding muscles, increased signal intensity on T2-weighted (T2W) MRI sequences, and heterogeneous enhancement following intravenous contrast administration. The mass extended along the dorsal aspect of the upper thoracic spine from C7-T1 to the inferior portion of T2, measuring 3.7 cm in craniocaudal extent, 1.7 cm in anteroposterior dimension, and 3.8 cm in transverse width. The adjacent thoracic spinal cord demonstrated anterior displacement and slight compression with obliteration of subarachnoid space posterior to the thoracic spinal cord. The mass extended beyond the laminectomy site resulting in infiltration of the adjacent left paraspinal musculature (Fig. 1). The patient was scheduled for surgery for a resection of the presumed recurrent meningioma. Histopathologic analysis of the surgical specimen revealed desmoid fibromatosis. In retrospect, preoperative dynamic contrast-enhanced MR imaging (DCE-MRI) provided sufficient information to suggest an alternative diagnosis to meningioma.

Treatment

A T1-T2 laminectomy was performed, followed by bilateral paraspinal muscle flaps and bilateral trapezius muscle flaps. A complete surgical resection of the tumor was achieved. An intra-operative ultrasound of the dura showed a slight uniform thickening without intradural nodularity, so the decision was made not to open the dura. There were no postsurgical complications. Postoperatively, the patient reported impaired coordination and continued weakness of the left lower extremity and is currently recovering. The possibility of postsurgical radiation therapy was dismissed due to proximity to the spinal cord and the recurring and local reactive nature of desmoids.

Discussion

Desmoid fibromatosis is a mesenchymal neoplasm that arises from fascia, aponeuroses, and muscle connective

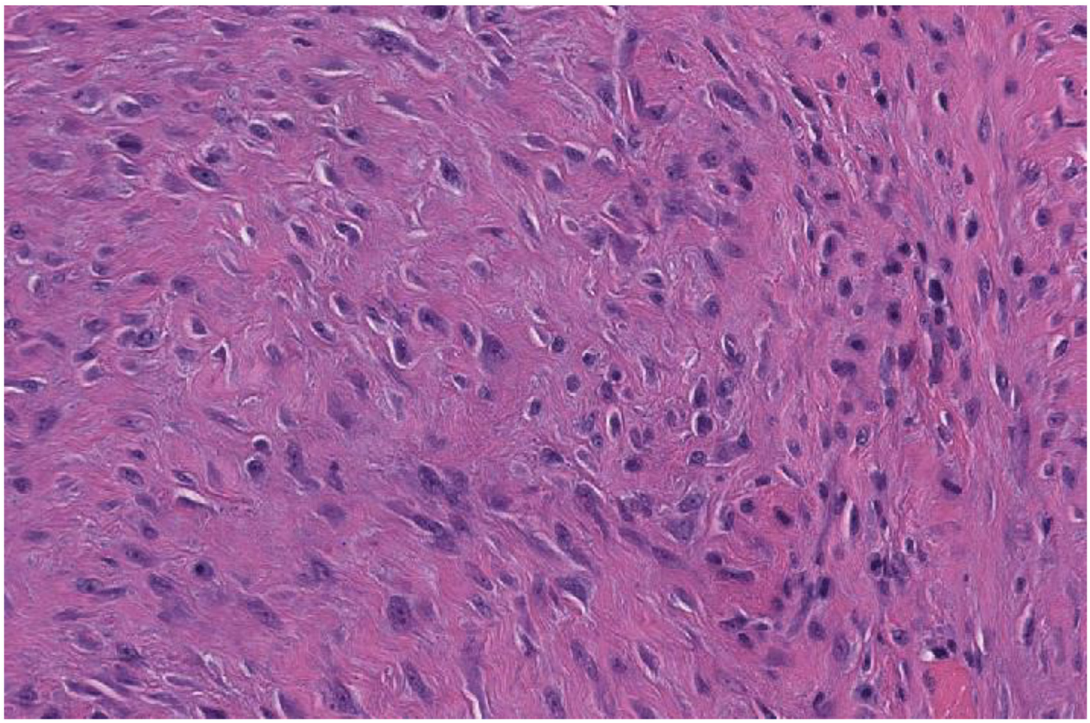

Fig. 3. – Photomicrograph of the excised tumor. The mass was identified as desmoid fibromatosis involving dense fibrous connective tissue with occasional calcification and atrophic skeletal muscle. The sample was heterogeneously described as firm, beige, and paucicellular with a predominance of collagen. In addition, spindle-cell neoplasms were identified microscopically.

tissue [3,5,13]. Although benign, it is locally aggressive and can infiltrate and compress the adjacent neurovascular structures causing pain and, rarely, life-threatening complications [2]. Most patients present with nonspecific symptoms like swelling and/or pain [2]. MRI is widely regarded as the imaging modality of choice for assessing the size of these masses and their interconnectivity with the surrounding structures, yet it remains difficult to accurately differentiate desmoid tumors from soft-tissue sarcomas using conventional MRI and other imaging techniques [14,15]. Desmoid tumors characteristically show low/similar signal intensity relative to the surrounding muscles on T1-weighted (T1W) images and variable intensity on T2-weighted (T2W) images [16]. Low-signal fibrous bands on T2W-MRI within a lesion with infiltrative margins is highly suggestive of the diagnosis.

DCE-MRI can improve specificity in the diagnosis of soft tissue tumors compared to conventional MRI. Using mathematical modeling, perfusion or "leakiness" (a combined representation of tumor blood flow, blood volume, and permeability) is quantified as a forward transfer constant (K^{trans})(17). High K^{trans} values correlates with increased angiogenesis and microvascularity; characteristic features of more malignant and aggressive types of neoplasms [17,18]. Desmoid tumors typically exhibit low tumor perfusion, which manifests as low K^{trans} values on DCE-MRI [17]. Meningiomas, on the other hand, are highly vascular lesions with higher K^{trans} values, more easily discernable from normal brain cortex and other less-vascularized tissues [19]. In this patient, K^{trans} value of the lesion appears equivalent to the intensity of the vertebral bodies (K^{trans} = 0.0289 min^{-1}), indicative of minimal leakiness and vascularity (Fig. 2a). In comparison, DCE-MRI of a known intracranial WHO grade II meningioma shows much higher K^{trans} value that is easily discernable from bone (Fig. 2b). Therefore, this imaging feature on DCE-MRI minimized the possibility of the mass being a recurrent meningioma (Fig. 3).

Historically, wide excision has been the mainstay treatment for desmoid fibromatosis. However, there has been a recent shift towards radiation and even chemotherapy-based treatment plans [20]. A consensus has yet to be reached for the most appropriate treatment, and remains dependent on the specifics of each case including tumor location, patient age, and past medical history. Following surgical resection of the presumed recurrent spinal meningioma, the current treatment strategy in this patient is regular follow-up and monitoring the now-identified extra-abdominal epidural desmoid tumor.

Conclusion

Post-traumatic desmoid fibromatosis, although rare, should be considered in the differential diagnosis of a mass recurrence at a postoperative site. DCE-MRI can characterize the perfusion features of lesions. The slow perfusion of desmoid fibromatosis, compared to lesions with high perfusion such as meningioma, can suggest an alternative diagnosis to

recurrent tumor and allow for optimal surgical planning. To our knowledge, we report the second case of desmoid fibromatosis occurring secondary to spinal meningioma where DCE-MRI features can help differentiating desmoid tumors from recurrent disease, even prior to biopsy or intervention.

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