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PRS GLOBAL OPEN CASE REPORT

Hand/Peripheral Nerve

Brachial Plexus Desmoid Tumor: Care for Functional Preservation

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Summary: Desmoid tumors are a rare, locally invasive, non-metastasizing tumor of mesenchymal origin. Most of such tumors occur sporadically, but some arise as part of germline adenomatous polyposis coli mutations. They tend to aggregate in the abdomen, thorax, extremities, and the head and neck region. They are challenging to treat, with a high rate of recurrence even if achieving negative margins. We present the case of an 18-year-old woman with a desmoid tumor involving her brachial plexus. A non-oncological resection was performed, with a focus on functional preservation. Residual disease is being treated with β-catenin inhibitor and monitored with serial MRI. *(Plast Reconstr Surg Glob Open 2020;8:e3293; doi: 10.1097/GOX.0000000000003293; Published online 21 December 2020.)*

Desmoid tumors account for 3% of all soft tissue tumors, with 900 cases reported annually in the United States.[1] Extrabdominal desmoids commonly arise in the extremities and, to a lesser extent, in the head and neck region (7%–15%).[2] Sporadic tumors tend to have an error in β-catenin, a proto-oncogene, which normally functions to regulate cell adhesion and cell transcription.[1,3]

CASE PRESENTATION

An 18-year-old woman was referred to our peripheral nerve surgery clinic for concern of a right brachial plexus sheath tumor. Three years before, she developed right shoulder pain and instability, and then noticed a slowly enlarging mass in her supraclavicular region. MRI indicated a mass (1.8 × 1.9 × 5.4 cm) between the right anterior and middle scalene muscles, running parallel to the course of the right C5/C6 nerve roots and upper trunk of the brachial plexus (Fig. 1). The radiographic impression was a nerve sheath tumor likely of schwannoma origin, possible neurofibroma or malignant peripheral nerve sheath tumor.

The patient reported worsening pain, instability, and electric-like shocks radiating from the deltopectoral region down her right lateral and dorsal forearm. Her right upper extremity strength was preserved with the exception of minor shoulder weakness and instability.

We performed an excisional biopsy using the standard supraclavicular brachial plexus approach. Intraoperatively, a considerable amount of fibrous tissue was noted surrounding the upper, middle, and lower trunks of the brachial plexus (Fig. 2). The upper trunk was traced back to C5/C6 nerve roots; however, there was no abnormality of the roots, as noted on MRI. Posterior to the C5 nerve root, within the middle scalene muscle, was a large and firm palpable mass. A nerve was seen entering the mass proximally. Stimulation of this nerve contracted the serratus anterior muscle, and this nerve was identified as the long thoracic nerve.

At this point, concern was raised for a possible soft tissue malignancy or a malignant nerve tumor. Intraoperative frozen sections were sent and showed cytologically bland spindle-cell proliferation surrounding nerve sheath structures, without evidence of high-grade malignancy. Intraoperative consultation with surgical oncology recommended non-oncological resection with the priority of preserving upper extremity function.

Postoperatively, the patient had an uneventful recovery and reported resolution of her shoulder pain. Final pathology showed the mass to be a desmoid tumor, demonstrating cytologically bland myofibroblastic proliferation forming long, sweeping fascicles with nuclear expression of β-catenin and smooth muscle actin. Resection margin was positive. The patient was referred to a multidisciplinary oncology clinic. Radiation therapy was discussed but not recommended due to risk of radiation-induced perineural fibrosis. The patient is currently enrolled in a clinical trial with β-catenin inhibitor (Fig. 3). At 6 months postoperatively, repeated MRI showed a residual tumor

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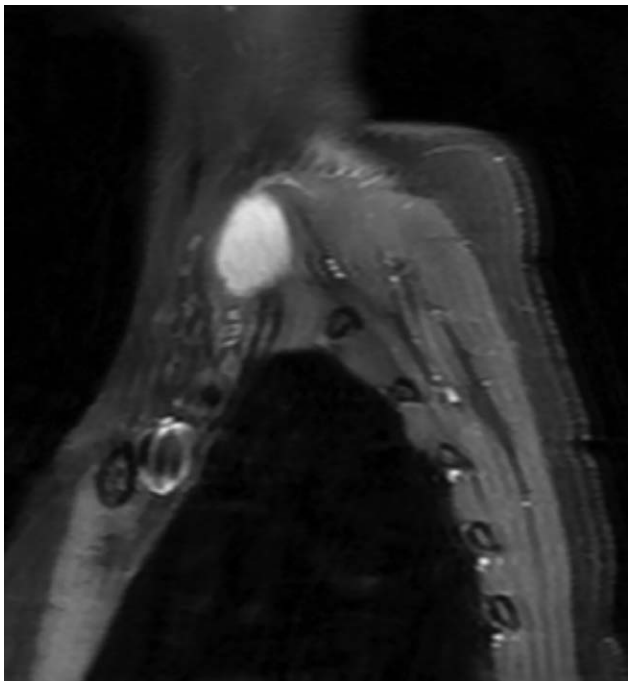

Fig. 1. MRI demonstrates a large mass around the right brachial plexus.

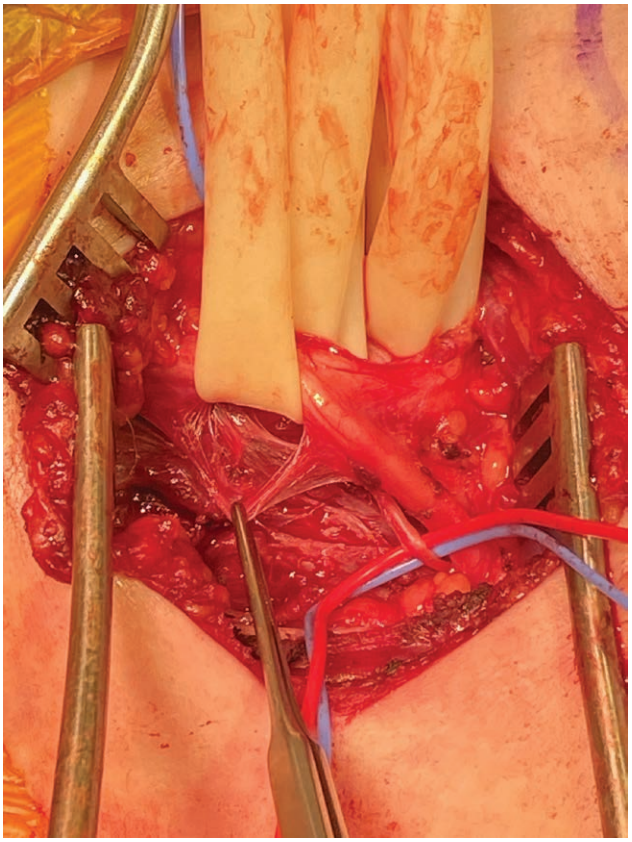

Fig. 2. Intraoperative photograph showing the brachial plexus encased by significant fibrous tissue.

with a slight increase in size (Fig. 4). Her right upper extremity currently has normal motor function, and she is employed full time.

DISCUSSION

As desmoid tumors are rare, benign, and with no metastatic potential, there is controversy as to the optimal treatment.[2,3] Although resection with clear margins is ideal, there are still cases with local recurrence after a wide local excision. Conversely, there are also cases of positive margins without subsequent disease progression.[1,2] Over the last decade, treatment has shifted toward a "wait-and-see" approach focusing on conservative care.[1,3,4] Fiore et al. showed that a wait-and-see approach had comparable rates of progression-free survival at the 5-year mark, when compared with medical therapy, including hormonal therapy, low-dose chemotherapy, NSAIDs, and imatinibed mesylate.[5] However, roughly 50% of patients in either group had progression of their tumor, suggesting that surgery should be reserved for aggressive cases.[1,2,6] While margin status is usually of utmost importance for surgical resection, desmoid tumors challenge this ideology, as margin status does not provide prognostic value in the development of disease.[1,2,7]

Of the extra-abdominal fibromatoses, only 12% arise in the head and neck region, and this location may be more aggressive than the abdominal wall possibly due to restricted anatomy, vital vasculature, and neural structures.[2,8] Of the head and neck desmoid tumors, Kruse et al. found that neither age, sex, nor localization led to a difference in outcomes.[8] Hoos et al. found that only 9 of their 21 head and neck desmoid patients had full resection without involvement of surrounding structures, with 8 having good functional outcomes and 13 having persistent functional problems.[9] They concluded that due to uncertain rates of recurrence reduction with negative margins, the goal of surgery should be function-sparing, rather than complete resection.

Adjunctive radiotherapy is often added to unresectable desmoids or those with positive margins; however, complications such as tissue fibrosis, radiation-related cancer, and skin damage may be significant.[2] Hoos et al. conclude that there was not strong evidence to support the use of radiotherapy in the treatment of head and neck desmoids, regardless of margin status. Consequently, radiation-related morbidity must carefully be weighed against the benign nature of desmoids.[1,9] Despite the risks, radiotherapy following resection is still used in some cases, and may have the most utility in instances with recurring desmoids.[1,3,10]

Chemotherapy has been reserved for failure of surgery or radiation. However, some newer studies show an early response, achieving disease stabilization in two-thirds of patients.[1] This is an area of active research and may ultimately shift treatment guidelines for desmoid tumors. Gounder et al. examined the role of sorafenib as an adjuvant treatment, and found 16 of the 22 symptomatic patients had clinical improvement within 2 weeks.[4] Other medical therapies such as tamoxifen, NSAIDs, and various

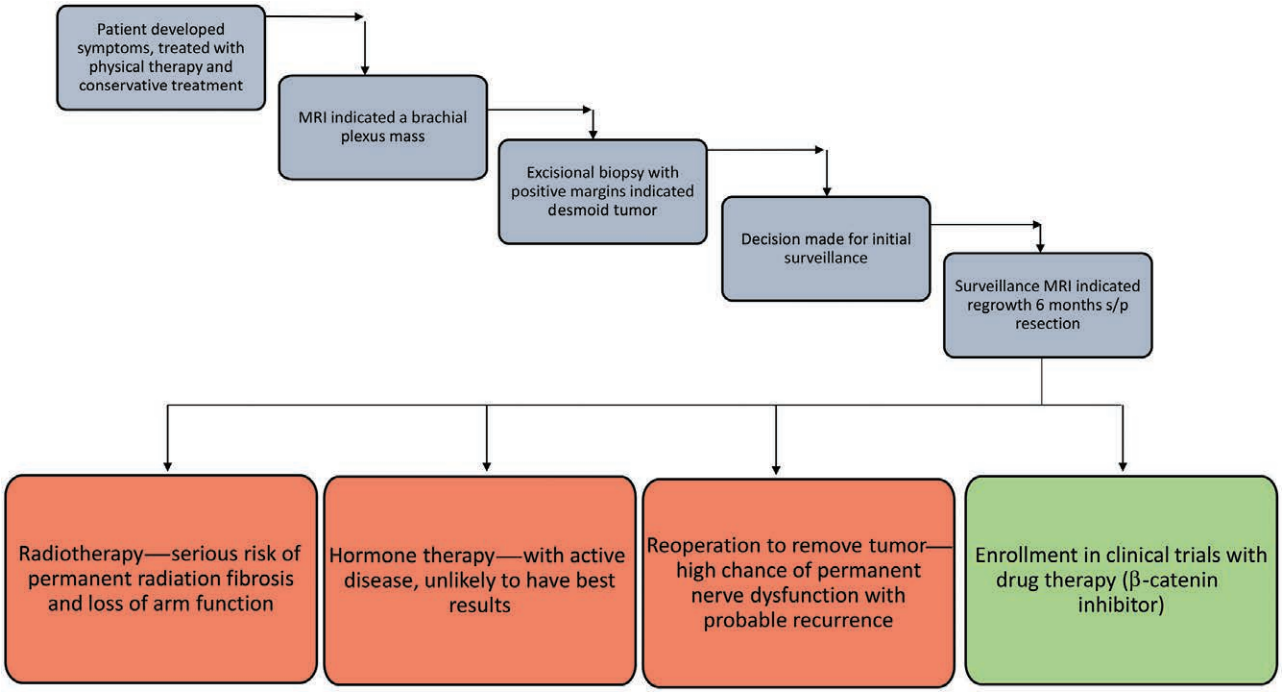

Fig. 3. Decision tree for desmoid tumor management in our patient.

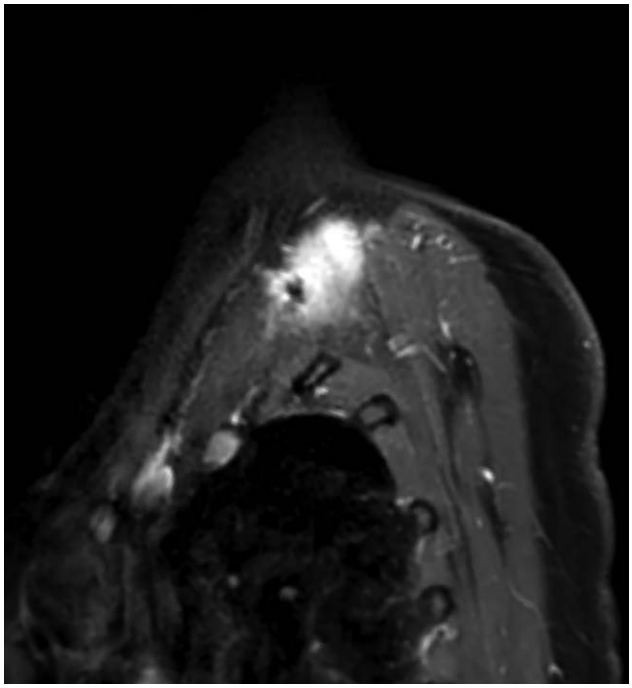

Fig. 4. Six months postoperative MRI showing residual tumor with regrowth.

chemotherapy regimens have been used, but there is no consensus on a preferred regimen.[3,4]

In our patient, surgical excision confirmed the diagnosis but, due to the proximity of the tumor to the brachial plexus and the significant risk of functional deficits should aggressive resection be attempted, decision was made by our multidisciplinary tumor board to prioritize medical management even with local recurrence. If she continues to worsen clinically, particularly if local progression causes upper extremity weakness or significant pain, we may consider operative intervention with repeat resection and nerve reconstruction.

CONCLUSIONS

Desmoid tumors are a rare and challenging disease to treat. Care needs to be taken to preserve function of involved structures because recurrence is common, even with negative margins. A conservative "wait-and-see" approach may lead to optimal patient outcomes, but a multidisciplinary team utilizing a spectrum of therapies is key, as tumors are likely to progress with no one single treatment option demonstrating a high efficacy.

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