Myofibroma of the cervical spine presenting as brachialgia

Case report

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Myofibromas are rare, benign tumors of myofibroblasts. Their occurrence in adults, involving bone outside of the head and neck, is especially uncommon. The authors report the case of a 34-year-old woman who presented with left-sided brachialgia. Magnetic resonance imaging identified an expansile soft-tissue lesion of the C6–7 facet joint. En bloc resection via a left posterior midline approach was undertaken. Histopathological analysis confirmed the lesion to be a myofibroma. Brachialgia resolved following surgery and there is no evidence of recurrence at 20 months follow-up. Myofibroma is a rare cause of primary soft-tissue tumor of the spine. Surgical excision remains the mainstay of treatment.

Key Words • myofibroma • cervical spine • brachialgia • oncology

Myofibromas are rare, benign tumors of myofibroblasts more commonly found in children than adults. These tumors typically involve soft tissue rather than bone, with a predilection for the head and neck. The involvement of bone may have an association with multicentric myofibroma, or “myofibromatosis.” As such, the occurrence of an extracranial, solitary, osseous myofibroma is especially uncommon. To our knowledge only 2 previous cases in the cervical spine have been reported. We describe the presentation, diagnosis, and management of a myofibroma arising from the C6–7 facet joint.

Case Report

History and Examination. A 34-year-old woman presented with a 10-month history of progressive neck pain and left-sided brachialgia. Her neurological examination was unremarkable and there was no relevant medical history.

Magnetic resonance imaging of the cervical spine revealed an expansile soft-tissue mass arising from the left C6–7 facet joint, with compromise of the exit foramen (Fig. 1A–C). Local bone erosion was also evident on CT (Fig. 1D). The associated remodeling of the medial cortex of the C-6 lamina suggested a degree of chronicity. There was peripheral enhancement with Gd administration (Fig. 1B), and overall the lesion remained well circumscribed.

Computed tomography of the thorax, abdomen, and pelvis failed to show any other lesions. A SPECT bone scan with 99mTc revealed no abnormal osteoblastic activity in relation to the C6–7 lesion or elsewhere.

A CT-guided biopsy was undertaken, which obtained a very scant amount of tissue revealing myofibroblastic-type cells on histological analysis. Rare mitotic figures were apparent. Overall, a benign soft-tissue tumor appeared more likely, but given the possibility of a low-grade sarcoma and the patient’s ongoing symptoms (i.e., brachialgia), surgical excision was deemed appropriate after multidisciplinary discussion.

Operation and Postoperative Course. Surgical excision was undertaken via a left-sided posterior midline approach. A fairly firm, avascular, and well-encapsulated lesion was noted to arise from the inferior portion of the left C6–7 facet joint, with well-defined margins. The lesion was easily dissected off the almost fully eroded left C-6 lamina and C6–7 facet joint laterally, and the attachment resected en bloc with an osteotome. Adjacent bone surfaces were comprehensively curetted and a left hemilaminectomy completed. There was no involvement of the

Abbreviation used in this paper: SMA = smooth muscle actin.
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adjacent dura or the exiting nerve root. We elected to not instrument the C6–7 level, given the possibility for further surgery (i.e., in case the histological analysis confirmed a sarcoma) and to minimize artifacts on postoperative MRI.

The patient made an uneventful recovery and reported resolution of her left-sided brachialgia. She remains well at 20 months follow-up and the postoperative MR image at 19 months continued to show no evidence of tumor recurrence or spinal deformity (Fig. 2).

**Pathological Analysis.** Pathological examination of the lesion revealed a tumor characterized by a distinctive pattern of bands of hyalinized collagen lacking elastin content and intersected by strands of pale spindle cells showing limited cytological atypia (Fig. 3A). Mitotic figures were exceptionally sparse and nuclear Ki-67 labeling less than 1%. Small, normal-appearing capillaries were associated with some of these strands of tumor cells (Fig. 3C). The tumor cells were immunoreactive for vimentin and smooth muscle actin (SMA; Fig. 3B) with negative staining for AE1/3, CD34, Bcl-2, MNF116, EMA, S100, desmin, and CD99. The appearances were considered consistent with a sclerosed or desmoplastic myofibroma of bone.

**Discussion**

A myofibroma is a rare, benign tumor of myofibroblasts, which are spindle-shaped cells of mesenchymal origin more synonymous with wound healing and granulation tissue. Their exact etiology remains unknown. It was first described by Stout in 1954, although the terms “solitary” and “multicentric” were coined in a series by Chung et al. in 1981.4,11

Myofibroma predominantly affects children, with a predilection for the soft tissues of the head and neck. In their series of 61 infantile cases, Chung et al. found that 88% arose in the first 2 years of life.4 More recently, a series of 114 cases including adults by Oudijk et al. in 2012 found that 65% presented in the first 2 years of life and 91% before the age of 18 years.9 Multicentric myofibromas are less common than the solitary form, although they are more closely associated with bone involvement.5 Cases of solitary osseous myofibromas have been described, the vast majority affecting the mandible.8

Two previous cases of myofibroma of the cervical spine have been described. The first by Asirvatham et al. in 1994 arose from the odontoid peg, causing atlanto-occipital subluxation and neck pain in an 18-year-old man.3 The second case by Swierkowski and Seex in 2004 arose from the C7–T1 intervertebral foramen of a 48-year-old man, presenting with brachialgia.12 In the latter case, resection of the lesion through a laminoforaminotomy revealed that the tumor was arising from the bone within the foramen and separate from the exiting nerve root. These findings are similar to that observed in the present case.

Radiography and CT may identify a radiolucent, well-demarcated lesion with peripheral sclerosis. The MRI findings are usually of moderate signal on T1-weighted imaging, high signal on T2 weighted images, and with peripheral enhancement. However, the radiological appearances are not specific and similar appearances may be observed with metastases, myeloma, and osteoblastoma.8 A tissue sample is therefore the key to diagnosis. Macroscopically, myofibromas appear as well-demarcated nodules of variable color.8,9 Microscopically, myofibroblasts appear spindle-shaped, with elongated nuclei and a pale eosinophilic cytoplasm, features shared by fibroblasts, smooth muscle cells, and pericytes.9 Such similarities can cloud diagnosis, particularly on biopsy and in unusual locations. Allon et al., in a 2007 review of mandibular myofibroma, reported a 26% rate of misdiagnosis on biopsy, including spindle cell lesions of nerve tissue, smooth muscle aberrations, desmoplastic fibroma, and fibrosarcoma.7

Tissue architecture may offer greater distinction: bundles of spindle cells arranged around vascular spaces, simulating hemangioendothelioma, are typical.7 An abundant extracellular matrix is a feature of myofibroblastic tumors. Some examples such as the tumor under discussion are characterized by keloid-like stroma, referred to as the...
sclerosed or desmoplastic variant of myofibroma. Mitotic figures are uncommon and features of atypia are always absent, as observed in the present case. Immunohistochemical analysis is also helpful. Fibroblastic pathology can be excluded in the presence of strong immunoreactivity for actin. Stains for desmin and S100 are normally negative. Desmin reactivity becomes more likely in larger tumors. The behavior of myofibroma may differ depending on the tissue involved. A case-controlled series in 2012 by Abramowicz et al. identified soft-tissue myofibroma as more locally aggressive than intraosseous myofibroma. It is interesting to note that the CT features of our case suggested an element of chronicity.

Surgery is the mainstay of treatment, although descriptions of spontaneous regression do exist. No large case series has reviewed surgical strategies. Abramowicz et al. favored en bloc resection for exophytic myofibroma and enucleation for intraosseous myofibroma, with good outcomes. The choice of surgical technique may be complicated by diagnostic ambiguity, notably over concerns of a possible malignancy. Across the literature, en bloc resection does predominate as the primary treatment option. Recurrence is estimated at 10% and as such, long-term clinical and radiological follow-up is mandatory.

In summary, as an uncommon lesion among adults, myofibroma of the cervical spine is especially unusual. Radiology can locate and define the margins of the lesion, but histological analysis is required for a definitive diagnosis.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References


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