

Inflammatory Myofibroblastic Tumor of the Cervical Spine Mimicking Schwannoma: Case Report

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Abstract

Intraspinal inflammatory myofibroblastic tumor has been reported several times before. We are reporting one of the unique cases which are located in obliquus capitis inferior muscle and extending to cervical intraspinal epidural space. We present here 28-year-old female patient with a five months history of pain and parasthesia of right occipital area radiating to the same side of the neck anteriorly. Examination showed only reduced sensation of second cervical nerve root distribution. Computer tomography and magnetic resonance imaging showed contrast enhanced lesion inside obliquus capitis inferior muscle extending to cervical epidural spinal space between C1 and C2 vertebrae mimicking the appearance of dumbbell shape schwannoma. Gross total resection was done and histopathological examination showed inflammatory myofibroblastic tumor. Post-operative, her pain was completely resolved but numbness remained. Three years later, on the follow-up examination, she was pregnant and after the delivery of her baby magnetic resonance imaging showed complete resolution of the lesion. We are reporting this case here because of the both unique location and complete resolution of the inflammatory myofibroblastic tumor after pregnancy.

Keywords: Inflammatory myofibroblastic tumor; Extradural; Schwannoma; Pregnancy

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Introduction

Inflammatory myofibroblastic tumors (IMT) are soft-tissue masses of unidentified etiology, which has been described at various locations throughout the body. These lesions mostly affect the lung. The extrapulmonary localizations include the orbit, nasal sinuses, liver, spleen, pancreas, bowel, kidney, urinary bladder, testis, heart, lymphatic system, muscles and skin [1]. They rarely affect the spine [2]. Surgical excision is highly indicated to alleviate symptoms and to reach histopathological diagnosis. In some of the cases radiotherapy, chemotherapy and steroids are given as an adjuvant therapy. To the best of our knowledge, only seven cases of extradural inflammatory myofibroblastic tumor of the spine have been reported in the literature [2-7].

We present here an extremely rare location of inflammatory myofibroblastic tumor in the obliquus capitis inferior muscle between the levels of C1-C2 mimicking dumbbell-shaped schwannoma. After the surgical treatment complete resolution of the IMT is observed and did not show any recurrence even

after the pregnancy. This unique case is discussed with relevant literature.

Case Report

A 28-year-old female patient was admitted giving a five-month history of pain located in her right occipital area. She had paresthesia and mild restriction of neck movement. She did not mention any history of fever, weight loss, and contact with a patient with tuberculosis, ingestion of raw milk or trauma. On her neurological examination, she had decreased sensation in right occipital area throughout C2 dermatome. On her laboratory investigation, hemoglobin level, white blood cell count, erythrocyte sedimentation rate, C-reactive protein level, and results of liver function tests were in normal range. Chest X-ray did not show any abnormalities.

Cervical spine computerized tomography (CT) showed dumbbell-shaped isodense mass lesion in diameters of 3.4 × 3.6 × 2.7 cm

located in right obliquus capitis inferior muscle and extending into the epidural space between the level of C1 and C2 without bony involvement. After contrast injection this mass lesion was slightly enhanced in homogenously (**Figure 1 A-B**).

Magnetic resonance imaging (MRI) of the cervical spine showed the same dumbbell-shape mass lesion in the right higher cervical location. It was isointense to the spinal cord in T1 and hyperintense in T2-images. It showed in-homogenous enhancement after the gadolinium contrast. This lesion was showing indentation from the right epidural space into the cervical subarachnoid space leaving spinal cord intact (**Figure 2 A-D**).

She underwent operation because of both to relieve pain and to reach the tissue diagnosis. Hemi-laminectomy was done in the second cervical vertebrae and the extra-spinal part of the mass invading muscle was almost totally removed. Second cervical nerve root was sacrificed because of the tumoral invasion of the root.

Histopathological examination revealed proliferative spindle myofibroblastic cells with focal inflammation rich in plasma cells (**Figure 3**). Immune staining showed negative reaction to S-100 protein excluding neural origin and positive for vimentin confirming mesenchymal origin and positive for small muscle actin. This excludes schwannoma and confirms the diagnosis of IMT.

Follow up CT scan showed gross total removal of the mass. The patient symptoms were completely resolved except numbness and returned back to her previous work as a physiotherapist. One year later MRI cervical spine showed residual tumor as shown in **Figure 4**. Three years later she became pregnant and her follow-up MRI after giving a birth showed that spine is free of tumor and no recurrence (**Figure 5**).

Discussion

Several synonyms including fibrous xanthoma, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, benign myofibroblastoma, inflammatory fibrosarcoma, xanthoma, histiocytoma, xanthogranuloma, postinflammatory tumor, and inflammatory pseudotumor are the names given to describe inflammatory pseudotumor since it is first described in 1954 [8]. It is a soft tissue lesion of unknown pathogenesis. The first case of intraspinal cervical IMT was reported by Eimoto et al. [9] in 1978. In 2002, the World Health Organization accepts the term inflammatory myofibroblastic tumor (IMT) [10]. In most documented cases of IMT, the etiology remains unknown.

IMTs are important because of the difficulty in differentiating them from true neoplasm clinically, radiologically and intraoperatively [11]. *CT scan showed only soft tissue mass but no bony involvement* and this goes with other reports where they found no bony involvement [12]. MRI findings were iso-intense in T1, hyper-intense in T2 and homogenously enhancing after contrast. The larger part of the tumor was paraspinal destructing right obliquus capitis inferior muscle and extending between C1/2 to involve the spinal canal in the epidural space giving the appearance of dumbbell shape schwannoma of C2 nerve root but without cord compression symptoms.

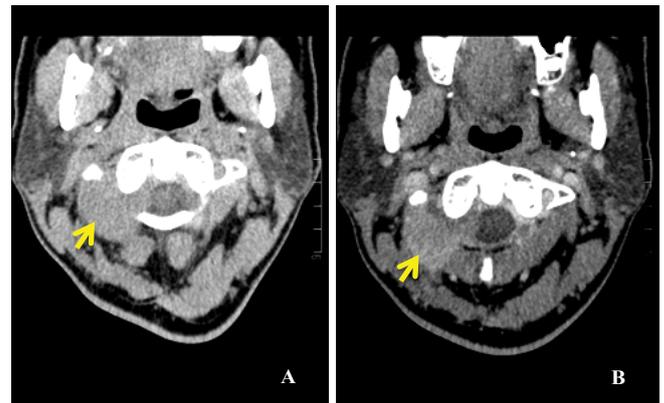


Figure 1 A-B: CT left without and right with contrast showed enhancement.

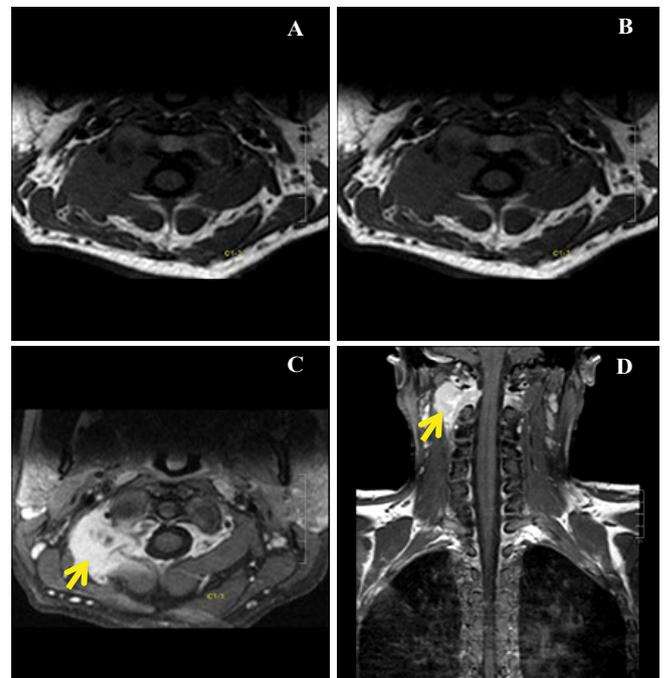


Figure 2 A-D: Cervical spine MRI showed right dumbbell shape lesion extending from paraspinal to intraspinal extradural space. Isointense in axial T1W1 (a) hyperintense in axial T2W1 (b) compared to the spinal cord. (c) after gadolinium tumor showed strong inhomogeneous enhancement. (d) Coronal T1W1 with contrast.

There are some reports referred to the origin of IMT from muscles. Paraspinal muscles in dorsal spine associated with rib fracture [13]. Isolated muscle involvement was reported in the anterior abdominal wall muscles [14,15]. In our patient the mass involved and infiltrated the obliquus capitis inferior muscle.

It has been reported involving various organs increasingly but intraspinal involvement of IMT is still very rare [2]. Intraspinal localization of the lesion can be primary or occur from a pulmonary IMT extension to the neighboring vertebrae [16]. Our patient presents with radicular symptoms involving C2 nerve root due to compression of this root in exiting foramen. This is the first case of IMT that mimic dumbbell schwannoma. Tumours with

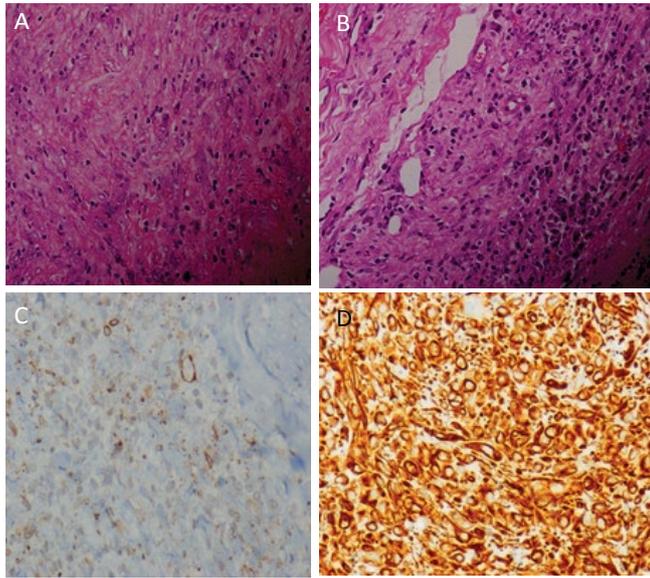


Figure 3 Photomicrographic view: lower left SMA positive and right lower vimentin positive.

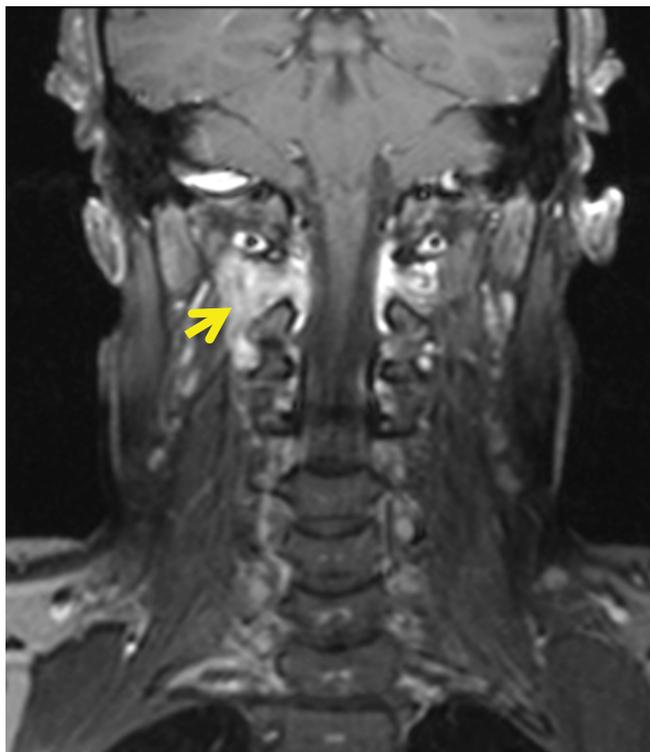


Figure 4 Residual tumor arrow head one year postoperatively.

both intraspinal and extraspinal extension, connected through the intervertebral foramen, are called ‘dumbbell’ or ‘hourglass’ tumours. Most dumbbell tumours are neurogenic tumours, including schwannoma, neurofibroma, ganglioneuroma, and neuroblastoma; schwannomas account for about 70-90% of all dumbbell tumours.

Ozawa et al. [15] in 2007 studied 674 spinal cord tumors. 118 (18%) were dumbbell tumors, schwannomas constitute 69%. Of

these 81 cases of schwannomas 18% were observed in the C-2 nerve root, thus having a higher incidence than those in the other nerve roots. These make us to consider our case preoperatively as a schwannoma. The rate of dumbbell tumors in the cervical spine was significantly higher than that of all spinal cord tumors. Malignant dumbbell shape tumors were found in 10 cases (8.5%) [16]. Spinal dumbbell-shaped schwannomas seem quite common, running to 10-15% of all spinal schwannomas [17-19]. Some researchers consider the dumbbell tumor as a typical shape for spinal schwannoma [16,18]. We found only 7 cases of extradural spinal IMT reported in literature (**Table 1**) [2-7]. None of these cases have dumbbell shape appearance.

Surgical excision is mandatory to relieve the symptoms and for cure and to reach histological diagnosis [20]. Our patient underwent surgical excision. Gross total resection for the paraspinal part and debulking of the epidural part. Although essentially considered benign lesions, IMTs may recur, metastasize or undergo malignant transformation [14]. Systemic steroids, radiation therapy and immunosuppressive drugs are also given in IMT in other sites and lead to decrease in volume of the mass [1,3,21,22]. Our patient was followed for more than 48

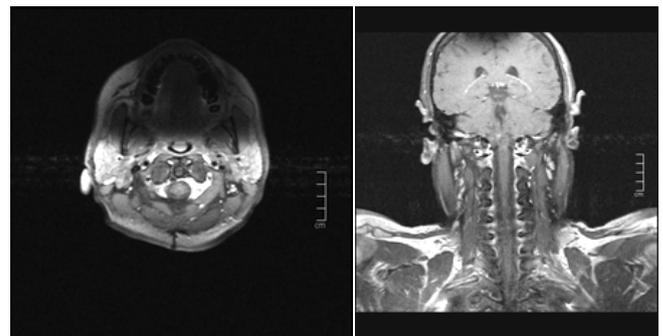


Figure 5 Four years after excision: axial and coronal contrast MRI showed no residual tumor at C1-C2 level.

Table 1 Summary of literature with spinal epidural IMT.

Refer No.	Year	Age (years)/ Sex	Location	T1W1	T2w1	Enhancement
Roberts et al.	1997	58/F	T9-T11	Iso	Hypo	NR
Gilliard et al.	2000	45/M	C3-T2	Iso	NR	Well
Roberts et al.	2001	39/F	T5-T6	Iso	Hypo	NR
Seol et al.	2005	44/M	T1-T7	Iso	Iso-Hyper	Well
Sailler et al.	2006	78/M	C6-T3	NR	Hypo	Well
		73/F	T5-T7	NR	Hypo	Well
Kato et al.	2012	63/M	T5-T6	Iso	Hypo-Hyper	NR
Our case		28/F	C1-C2	Hypo	Hyper	Well

T1W1: weighted images, T2W1: weighted images, F: Female, M: Male, Iso: Isointensity, Hypo: Hypointensity, Hyper: Hyperintensity, NR: Not Reported.

months and the last MRI scan showed no evidence of recurrence. The recurrence rate varies from <2% for pulmonary lesions confined to 25% for extrapulmonary lesions [23,24].

Inflammatory myofibroblastic tumors were reported during pregnancy [1,25,26]. One present as an acute spinal cord compression [1] and another one as upper airway obstruction. It appeared that there is no association with pregnancy. Follow up MRI after pregnancy showed complete resolution of the tumor in our patient.

Immunohistochemistry; anaplastic lymphoma kinase (ALK) overexpression was associated with early multifocal recurrence [26]. This is a prognostic laboratory test. In our patient this test

was not performed. But after more than four years with no recurrence it appears that it will be negative.

As a conclusion, inflammatory myofibroblastic tumors simulate neoplasm clinically and radiologically. Epidural IMT is rare but should be kept in the differential diagnosis of dumbbell shape schwannoma. Excision is mandatory for cure and histology diagnostic. Due to the limited number of reported cases, further research is necessary to better evaluate the efficacy of various treatment modalities. This is the unique case reported both in terms of its location originating from the musculus obliquus capitis and mimicking dumbbell–shape schwannoma and long-term follow-up during pregnancy.

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