Format for ANSWERING REVIEWERS

August 8, 2013

Dear Editor,

Please find enclosed the edited manuscript in Word format (file name: 3766-review.doc).

Title: Low Grade Spinal Malignant Triton Tumor with Mature Skeletal Muscle Differentiation

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Name of Journal: *World Journal of Neurology*

ESPS Manuscript NO: 3766

The manuscript has been improved according to the suggestions of reviewers:
1 Format has been updated

2 Revision has been made according to the suggestions of the reviewer

(1) Comments: This is a nice case report of a rare low-grade sarcoma in the extradural region of C2-C4. The manuscript is well written and the Figures are clear. The only comment is that it would be interesting to present data of the general medical examination, the medical history and current medication of this patient.

Response: Per the reviewer suggestion, more clinical symptoms and physical examination findings were included in the manuscript, clinical history section. A 53-year-old female without family history or stigmata of neurofibromatosis presented with worsening neck pain and right sided neck stiffness. The pain is present in the left side (buzzing in left hip and left heel). The quality of the pain is described as aching and shooting (pulling, popping) and the severity of the pain is at 6/10. The symptoms are aggravated by position and twisting. The pain is worse during the night while the stiffness is present all day. Physical examination is unremarkable except for right facial nerve weakness nerve. MRI showed a homogenously enhancing C2-C4 intraspinal extradural lesion exiting out the foramen at C2-C3 in a dumbbell fashion (Figure 1 A , B). A C2-C4 laminectomy was performed by using a combination of Leksell rongerus, Kerrisons and curettes. A tannish shear mass was identified in the epidural space from C2 to the top of C4. The mass was debulked from the inside using the combination of pituitary rongeurs as well as CUSA debulking and bipolar electrocautery. The post-operative MRI showed markedly decreased mass effect on the cervical cord (Figure 1 C). Bone stimulator and cervical collar were placed after surgery. The patient is treated with Percocet for analgesia and managed by physical therapy. No adjuvant treatment no radiation treatment were started. She is currently under regular follow up every six month (Figure 1 D, 10 months post-operative MRI), without evidence of recurrence (23 months post surgery).

(2) Comments: This descriptive case report outlines a clinical case of low-grade malignant triton tumor with unusual location and distinctive mature skeletal muscle differentiation. This Reviewer suggests that the inclusion of the measurements of the other myogenic regulatory factors (MRFs) would provide further information about the skeletal muscle component. In addition, this Reviewer suggests include more discussion on the negative immunostaining results of Myogenin and MyoD.

Response: We very appreciate the review's comments. 1) MyoD1 and myogenin are members of the Myogenic transcriptional regulatory protein, are the most widely used markers to express the early in skeletal muscle differentiation. They are very sensitive and specific for the diagnosis of immature skeletal muscle in rhabdomyosarcoma. Myf-5 and MRF4 are expressed at specific limited periods in myogenesis. However, they have not been routinely used for the diagnosis of immature skeletal muscle differentiation clinically, which are also unavailable in our hospital immunohistochemical laboratory. 2) The discussion of the negative immunostaining results of Myogenin and MyoD is re-written in the revised manuscript: In contrast to conventional MTT which are positive for rhabdomyoblastic markers Myogenin and MyoD-1, both markers are negative in our case. MyoD1 and myogenin are members of the Myogenic transcriptional regulatory protein, are the most widely used markers to express the early in skeletal muscle differentiation. They are very sensitive and specific for the diagnosis of immature skeletal muscle in rhabdomyosarcoma. Mature skeletal muscle component in a diffusely infiltrating peripheral nerve sheath tumor with hypercellularity and nuclear atypia is very unusual. Although the clinical indication of such features is still undetermined, the mature skeletal muscle differentiation might indicate a better prognosis than the conventional MTT, evidenced by the prolonged disease free interval of our patient (23 month post surgery without chemotherapy and radiation therarpy).

3 References and typesetting were corrected

Thank you again for publishing our manuscript in the *World Journal of Neurology*.

Sincerely yours,

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