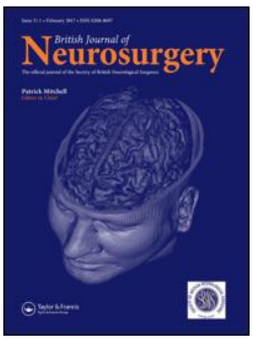

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SHORT REPORT

Collision tumor between a spinal schwannoma and chronic lymphocytic leukemia/small lymphocytic lymphoma: case report and review of the literature

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ABSTRACT

A collision tumor is one where two neoplasms of differing type occur at the same anatomical site. We present a patient suffering from non Hodgkin small cell lymphocytic lymphoma/chronic lymphocytic leukemia (SLL/CLL) and complaining intense lumbar back pain refractory to medical treatment. Lumbosacral MRI showed an intradural extramedullary lesion in the left L2-L3 foramen with extracanalar development and compression of psoas muscle. CT showed intralesional calcification. The patient underwent resection of the lesion through a paraspinal posterolateral approach (Wiltse approach). The histology was of schwannoma with intralesional calcifications and lymphocitic infiltrates compatible with B-lineage SLL/CLL. After the operation the patient suffer from left psoas muscle motor weakness (3/5 MRC). Because of hematological disease progression, she underwent 6 cycles of chemioterapy (Fludarabine, Cyclophosphamide, Rituximab). At a six-month follow-up no recurrence or residual tumor upon lumbosacral MR imaging was detectable and the left thigh flexion returned normal. To our knowledge, this is the first described case in the literature of collision tumor between a solitary spinal Schwannoma and SLL/CLL.

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KEYWORDS
Spinal cord schwannoma; spinal cord tumor; metastasis; neurinoma

Introduction

Collision tumors are a rare event where two distinct neoplastic entities, with different histological features, arise in the same anatomical area. The most frequent combination is between solid and hematological tumors.[1,2] We describe the first case of a collision tumor between a solitary spinal schwannoma and a small cell lymphocytic lymphoma/chronic lymphocytic leukemia (SLL/CLL).

Case presentation

A 61-year-old woman presented with a nine-month history axial lumbar back pain. No neurological deficits were noted on physical examination. One month prior she had been diagnosed with SLL/CLL. Lumbar MRI showed an enhancing lobulated intradural extramedullary lesion in the left L2-L3 foramen. This lesion was mostly in the extraforaminal area compressing the psoas muscle without infiltrating it (maximum diameters $5.8 \times 7 \times 7$ cm). The radiological picture was suggestive of a spinal schwannoma (Figure 1). CT depicted numerous intralesional calcifications (Figure 2).

Four months after presenting, the patient underwent surgical removal of the lumbar lesion through a paraspinal posterolateral approach (Wiltse). The mass seemed firmer than the surrounding tissue and had a cleavage plane from the adjacent muscle. The L2 root was sacrificed given its involvement. Post-op she had thigh flexion weakness 3/5 that improved over days. Histology found a fibrotic lesion with extensive areas of tissue necrosis and neuronal structures, along with a positive reaction to immunohistochemical markers such as S-100, neurofilaments and anti-GFAP antibodies. The anti-EMA antibody reaction was not found. Multiple foci of calcification containing small-cell lymphocytic proliferation foci compatible with B-type lymphocytes (CD20+, CD5+, CD23+) and a minimal presence of T lymphocytes (CD3+) were described. The final diagnosis was that of Schwannoma with degenerative changes and bony metaplasia with CLL/SLL infiltrates (Figure 3).

During follow-up CT showed disease progression and she had FCR chemotherapy in six cycles (Fludarabine 40 mg/m^2 + Cyclophosphamide 25mg/m^2 for three consecutively days and Rituximab 375 mg/m^2 for one day; 1 cycle = 28 days). At a six-month follow-up neurosurgical outpatient evaluation the left psoas muscle function had returned to normal. No recurrence or residual tumor upon MR imaging were found (Figure 4).

Discussion

Examples of collision tumors are described in many different locations with a clear prevalence in solid organs and in the carcinoma/sarcoma or carcinoma/lymphoma combination.[1,2] In a recent study by Himchak *et al.*, analyzing 100 cases of collision tumor in patients affected by Hematolymphoid Proliferative Disorders (HLPDs), the most commonly found associated non-hematological neoplasms were colon carcinoma (17%), breast carcinoma (15%), prostatic carcinoma (12%). CLL/SLL was the most frequently found HLPD (18%) followed by large-B-cell lymphoma (17%), follicular lymphoma (14%), marginal zone lymphoma (10%), acute myeloid leukemia (8%) and the classic Hodgkin lymphoma (5%).[3]

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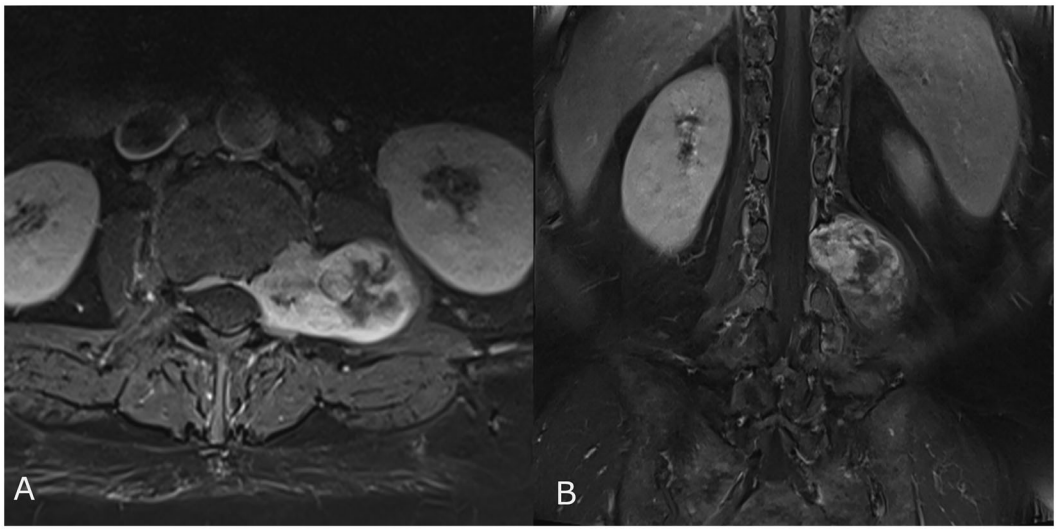

Figure 1. Pre-operative post-contrast T1 MRI revealing the large spinal schwannoma compressing and dislocating the ipsilateral psoas muscle: axial view (A) and coronal view (B).

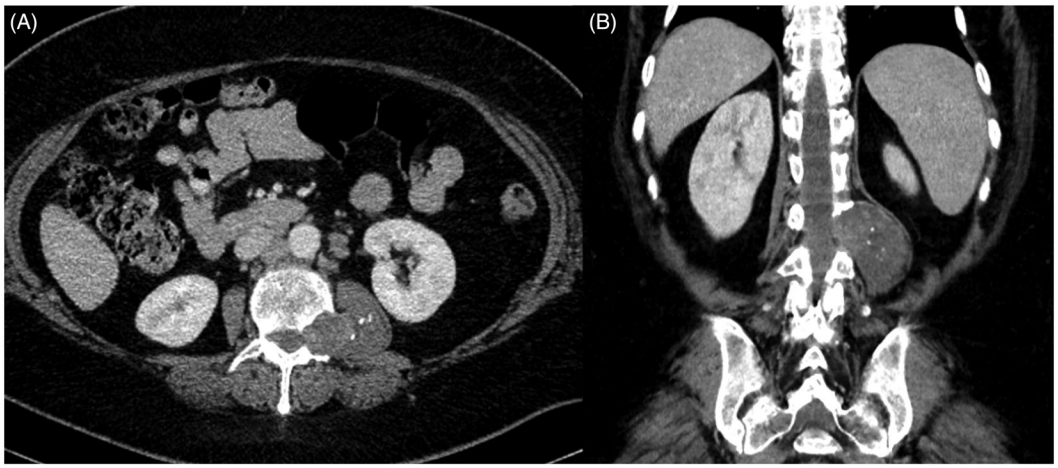

Figure 2. In Pre-operative abdominal CT scan revealing the multiple calcification within the spinal schwannoma: axial view (A) and coronal view (B).

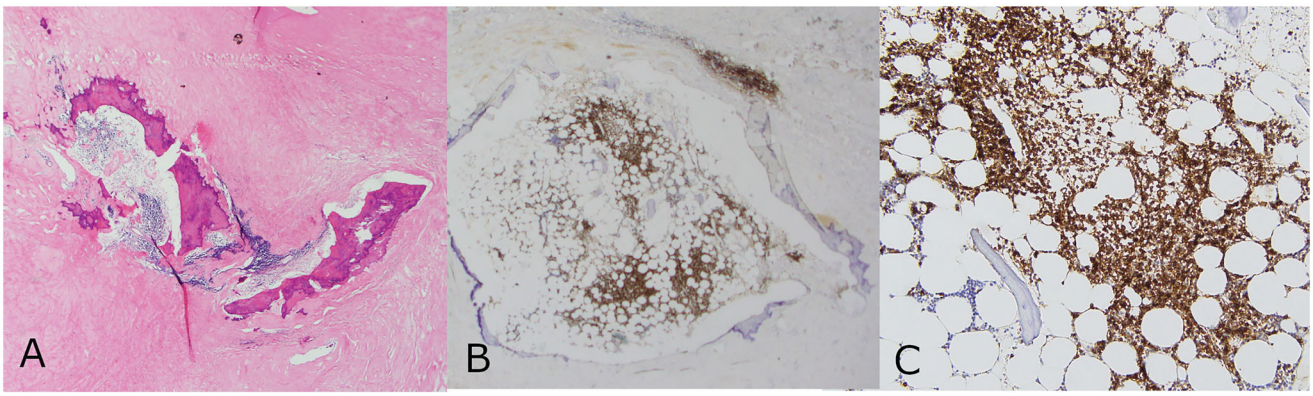

Figure 3. Photomicrographs of the resected lesion. Schwannoma with bony metaplasia areas (H&E ×100) (A). LLC/SLL proliferative foci located inside bony metaplasia areas within the Schwannoma tissue (100×) (B): the LLC/SLL foci show immunoreactivity for CD20 (Lymphocyte marker – 200×) (C).

Calcification within a schwannomas is well reported[4–6] but in our patient's case, these were not 'usual' bony metaplasia areas as they contained a monoclonal population of lymphocytic cells compatible with LLC/SLL.

In the literature only rare cases of collision tumors involving schwannomas are reported, with the exception of rare syndromes like Neurofibromatosis I and II.[7] The main combinations reported are with lung carcinoma, breast carcinoma and melanoma metastasis. Collision with hematological disorders are rare.[8]

However, anedoctal collision tumors between schwannomas and hematopoietic disorders of the lymphocytic T and B-cell lines have been reported.[9–11] The frequent presence of reactive

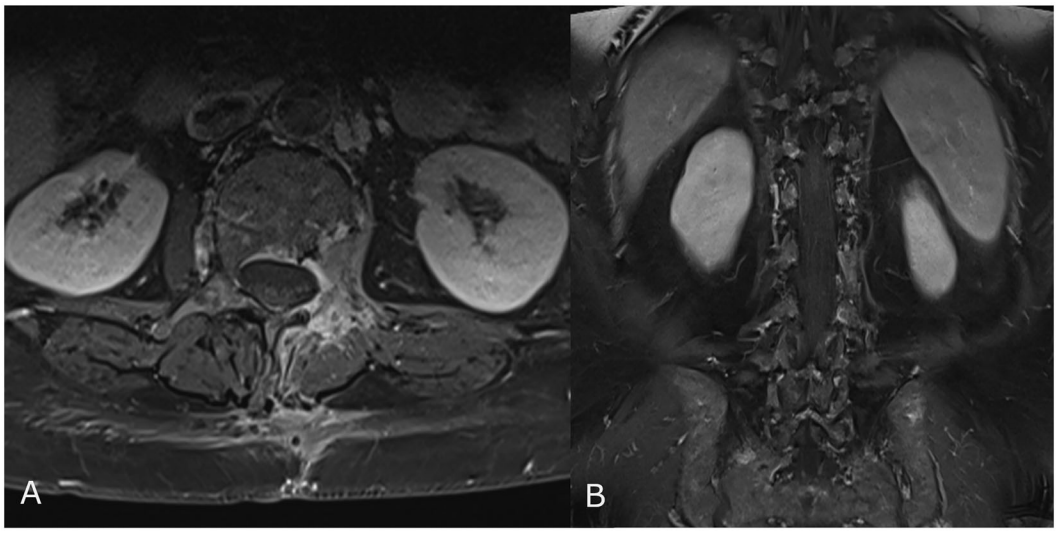

Figure 4. Post-operative post-contrast T1 MRI at 6 months follow-up revealing no residual and no recurrence of the tumor: axial view (A) and coronal view (B).

lymphoid infiltrates in the myelin nervous sheath complicates the correct diagnosis of collision tumor.

Two are the main theories regarding collision tumor pathogenesis. The first hypothesis, in the context of systemic illness, that carcinogenic stimuli could enhance the growth of two different tumors in within the same anatomical site.[11] The second focuses of tissue microenvironment changes: the favorable situation of "immunological escape" and increase of the local vascularization induced by the first tumor will facilitate the second tumor's growth.[11,12] Therefore a schwannoma, a slow well vascularized tumor (and thus not so competitive) could represent the optimal environment for the seeding and growth of other tumors, including the "low grade" types, as in our case.

Conclusion

To our knowledge this is the first case in the literature of collision tumor between a spinal solitary schwannoma and LLC/SLL.

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Informed consent

Written, informed consent was obtained from the patient for his information to be included in our manuscript. His information has been deidentified to the best of our ability to protect his privacy.

Disclosure statement

All authors have completed the ICMJE uniform disclosure form. The authors have no conflicts of interest to declare.

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