

## Case Report

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# Benign Collagen Rich Epithelioid Nerve Sheath Tumor in Cervical Spinal Cord: A Case Report

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## Abstract

Tumors that originate from the nerve sheath comprise diverse groups -perineuroma, neurofibroma and schwannoma. The epithelioid variant of these tumors is uncommon and mostly seen in malignant counterparts. Although this type of morphology is well recognized in peripheral nerve sheath tumors, its presence in the central nerves is rarely reported. Very few cases of nerve sheath tumors have been reported with collagen-rich stroma. Here we report an extremely rare case of nerve sheath tumor with epithelioid morphology and collagen rich stroma. To the best of our knowledge this finding in the spinal cord has not been reported thus far.

**Keywords:** Epithelioid collagen rich nerve sheath tumor, Cervical spinal cord

## Introduction

Nerve sheath tumors are a heterogeneous group of tumors that consist of schwannomas, neurofibromas and perineuromas.<sup>1</sup> Benign epithelioid nerve sheath tumors (BENST) are a group of tumors which are often diagnostically challenging because the epithelioid morphology in a nerve sheath tumor is most commonly indicative of a malignant rather than benign tumor.<sup>2</sup> Most reported cases of BENST (epithelioid schwannoma) have arisen from soft tissue and skin.<sup>3</sup>

Here we report our experience with a young lady who presented with cervical pain, which turned out to be a BENST. To the best of our knowledge this morphology in a nerve sheath tumor in the cervical spinal cord has not been reported thus far.

## Case report

A 36-year-old lady presented with long standing neck pain. Her main complaint since at least 6 months ago was cervical pain with no radiation. Her past medical history

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was completely unremarkable. She has two healthy children, with no specific familial history except for a lumbar laminectomy in her father performed several years ago.

Physical exam of the neck was remarkable for a non-radicular pain in the cervical to thoracic spine. Heart and lungs were completely normal. Both lower and upper extremity examinations were normal. No skin or subcutaneous mass, lesion or change in color was detected.

Laboratory results were normal for biochemical and hematologic analyses, with no abnormal tests. Molecular study for NF-1 showed the wild type.

Magnetic resonance imaging studies showed a well-demarcated encapsulated hyposignal T1 and hypersignal T2 space occupying lesion in the right side of the spinal process and posterior aspect of the C7 that measured 4 cm in greatest diameter (Figure 1).

The patient underwent surgery and an intradural extramedullary tumor was easily dissected and excised from the peripheral tissues. The pathology specimen was a well-defined encapsulated firm mass that had a creamy homogenous color. No necrosis or hemorrhage was detected.

Microscopic study of the mass showed a completely heterogeneous appearance. The bulk of the tumor was composed of epithelioid cells arranged mostly in sheets, with foci of rosette formation (Figure 2a). The tumor cells were bland in appearance with no atypia or mitosis (Figure 2b). There was no area of classic Antoni-A or B morphology in the examined sections. The specimen was totally embedded. The tumor cells were positive for S100 (Figure 2c) and vimentin, and negative for cytokeratin, epithelial membrane antigen (EMA), CD34, BC12, smooth muscle actin (SMA), melan-A and CD99. There was no necrosis or hemorrhage, but there were foci rich in collagen (Figure 2d). P53 was negative and Ki67 proliferative index was very low and less than 2% (Figure 2e). A diagnosis of BENST (collagen rich) or epithelioid schwannoma was made.

The patient spent an uneventful postoperative period and left the hospital in excellent general condition. After 3 months she was doing well

and completely symptom-free, with no complaints of neck pain.

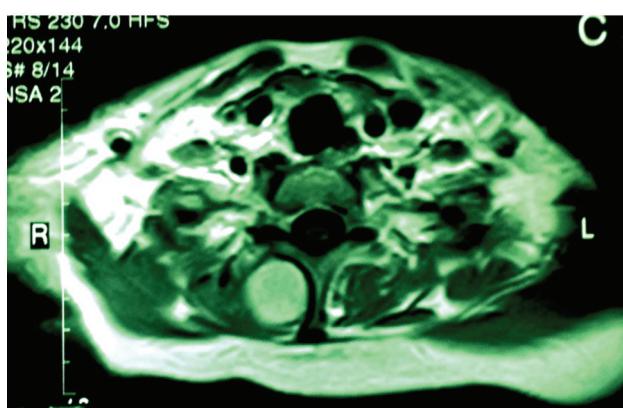
## Discussion

Nerve sheath tumors are a heterogeneous group of tumors which can be benign (benign nerve sheath tumor) or malignant (malignant nerve sheath tumor).<sup>3</sup> Epithelioid morphology, although rare, is most commonly seen in malignant cases and is indicative of aggressive behavior. This type of morphology is well recognized in peripheral nerves of the soft tissue and skin, however it is very uncommon in central and cranial nerves.<sup>4</sup>

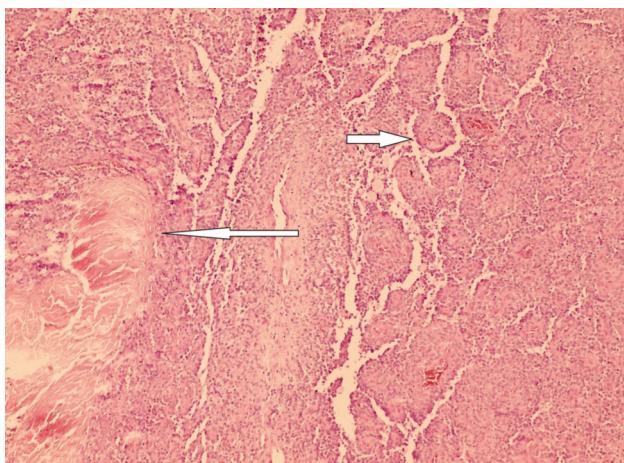
Epithelioid morphology in nerve sheath tumors of the skin, subcutaneous and soft tissue was reported in 33 cases by Laskinn et al. in 2005.<sup>3</sup> Epithelioid morphology has been rarely reported in the cranial nerves; there have been 4 cases reported thus far. In a report by Tan et al. in 2004, no evidence of malignancy in clinicopathologic investigations were found.<sup>4</sup>

The most challenging, important point for this type of tumor is the correct diagnosis and accurate exclusion of differential diagnoses. The gold standard for confirmation of the diagnosis in an epithelioid nerve sheath tumor is immunohistochemistry (IHC). Diffuse positivity for S100 is a very important diagnostic clue.<sup>3</sup>

In cases with collagen rich stroma reported by Jokinen et al.<sup>5</sup>, solitary fibrous tumor was a



**Figure 1.** Magnetic resonance imaging (MRI) showed a well-demarcated encapsulated hyposignal T1 and hypersignal T2 space occupying lesion in the right side of the spinous process and posterior aspect of C7.

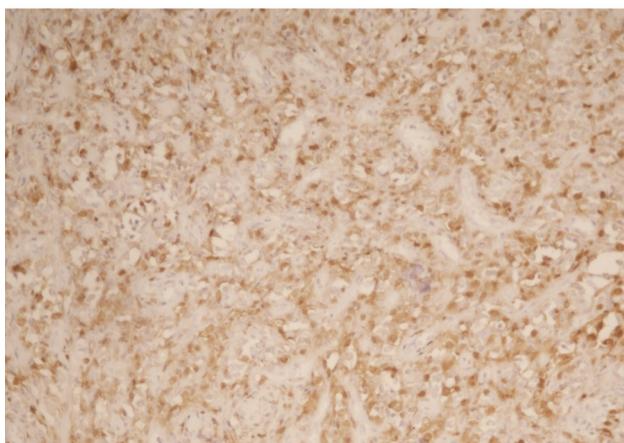


**Figure 2.** Heterogeneous histopathology of the tumor according to hematoxylin and eosin (H&E) and immunohistochemistry (IHC) results.; A. Sections from the tumor show rosette-like structures (short arrow) and collagen rich stroma (long arrow) with no atypia or mitosis (H&E; 250 $\times$ ) as well as epithelioid morphology.

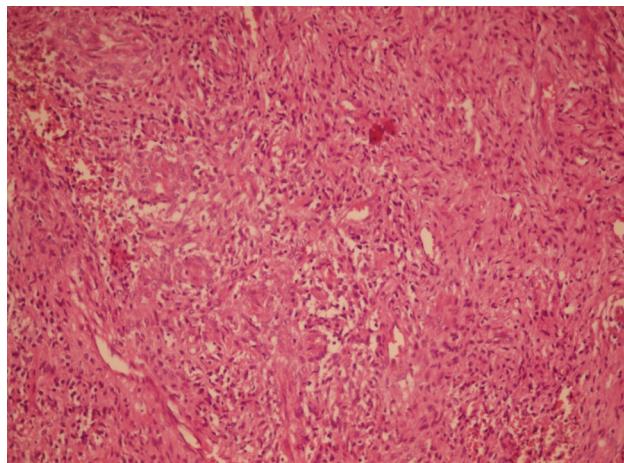
very important differential diagnosis, which could be excluded with nonreactive CD34 and reactive S100.<sup>5</sup> Our case showed areas with collagen rich stroma that resembled a solitary fibrous tumor, however this was excluded by IHC studies.

Other differentials such as meningioma and ependymoma are also excluded by an IHC study. Soft tissue tumors such as fibroma and leiomyoma are excluded by negative CD34 and SMA results.

Another important point in this case was the foci with rosette like morphology, which has been reported by Goldblum et al. in 1994 in an epithelioid schwannoma with collagen rich stroma, as with our case.<sup>6</sup> A number of authors have called this type of morphology a “neuroblastoma-like epithelioid schwannoma”.<sup>7</sup>



C. Reactive S100 in the nuclei of epithelioid cells.

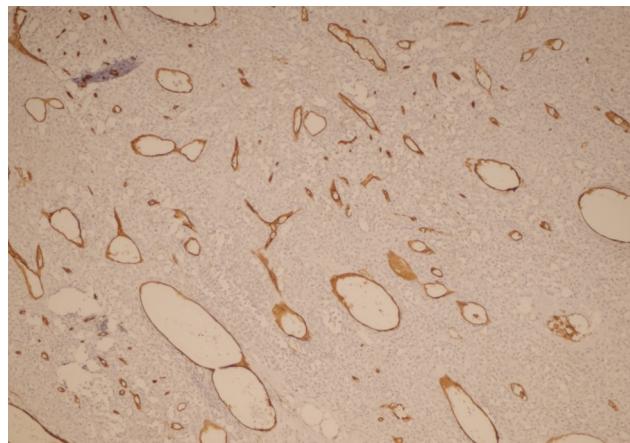


B. Areas of the tumor show spindle-shaped cells with no atypia or mitosis (H&E; 400 $\times$ ). Note that the stroma of the tumor is rich in collagen.

In a nerve sheath tumor with epithelioid morphology, malignancy should be diagnosed based on hematoxylin and eosin (H&E) slides, confirmed by IHC.<sup>8</sup>

Presence of atypia, mitosis and necrosis are strong evidences of malignancy which have not been found in our case. There should be a thorough examination of the tumor in order to locate any evidence of malignant behavior.<sup>8</sup> IHC for Ki67 is a reliable factor of the growth rate and possibility of aggressive behavior; also p53 positivity can be helpful in this regard.<sup>8</sup>

Our case showed epithelioid morphology, rosette-like structures, and foci of collagen rich stroma with no mitosis, atypia or necrosis. IHC showed a low proliferative index with Ki67 and negative P53. Follow up of our patient was unremarkable.



D. Non-reactive CD34.

In conclusion, nerve sheath tumors with epithelioid morphology are not always malignant and can be seen in peripheral, cranial and spinal nerve roots.

### **Conflict of Interest:**

No conflict of interest is declared.

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