CLINICAL VIGNETTE

Schwannoma of the Thoracic Esophagus

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Case Report

A 46-year-old Caucasian female presented to her Primary Care Physician for preoperative evaluation in anticipation of laminectomy. The patient has a history of chronic back pain, obesity, long-term tobacco use and obstructive sleep apnea. She reported gastroesophageal reflux symptoms and dysphagia and CXR noted a rounded mass in the right mediastinum, prompting computed tomography (CT) scan. This demonstrated a 3.0 by 2.7 cm mass within the esophageal lumen approximately 10.2 cm distal to the pharynx without surrounding mediastinal lymphadenopathy. The patient underwent esophagogastroduodenoscopy with fine needle biopsy of the tumor and the sample showed very faint DOG-1 focal staining. S100 also showed weak staining and the sample was negative for CD117, pan-cytokeratin, actin, desmin, and melan-A. Although this initial biopsy was inconclusive, the results did not support carcinoma, melanoma, or a smooth muscle malignancy and she was advised to have the mass excised. She underwent thoracotomy and resection of a 5.3 x 4.4 x 2.6 cm tumor, weighting 27.4 grams (Figure 1). Lymph nodes were negative for metastasis. An adherent 1.6 x 0.7 x 0.4 fragment of the esophagus was also resected and primarily repaired. She was kept NPO until tolerating clear liquids and was discharged home and is recovering well.

Final histologic sections of the excised esophageal mass show a submucosal proliferation of spindle cells with elongated nuclei with bland nuclear features (Figures 2, 3). There are numerous admixed reactive lymphoid follicles. Focally, there is a suggestion of Verocay body formation (Figure 4). Antoni A and B tissue types are also visible in these sections. Immunohistochemical stains demonstrate that these spindle cells stain with S100 (Figure 5) and SOX-10. Stains for CD34, CD117, Actin, AE1/AE3, Desmin, and DOG-1 are negative within the spindle cells. The negative reactivity for CD34 and CD117 rules out a gastrointestinal stromal tumor (GIST) and negativity for Actin and Desmin stains distinguishes the tumor from the more common leiomyoma. A stain for STAT6 is negative and a stain for neurofilament does not highlight axon within the lesion. These findings are diagnostic for Schwannoma, and there are no features to suggest malignancy.

Discussion

Esophageal schwannoma is an uncommon tumor that is rarely malignant. Literature review identified approximately 30 cases with only 4 neoplasms among those reported. Additionally, only two of these rare tumors had lymph node metastasis.[1]

This patient is only 46 years old, younger than the typical presentation, and is female, a significantly predominant selector. She is also Caucasian; one study reported 64% of these tumors were found among Asians.[2] These tumors typically present in the 5th to 6th decade in life with the youngest reported case in a 22 year old Asian male.[3] Although dysphagia (caused by compression of the esophagus) and dyspnea (caused by compression of the trachea) are frequent symptoms of this, and other, esophageal tumors, chest pain, stridor, hematemesis, and cough have also been reported.

Radiologically, these tumors are indistinguishable from other, more malignant esophageal tumors and diagnosis must be histologically confirmed. Histologic characteristics of these tumors include haphazardly arranged bundles of bland spindle cells with elongated nuclei and axons missing on neurofilament stains and a lack of necrosis or mitotic activity. Antoni A tissue demonstrates this palisading cellular arrangement and associated Verocay bodies reflecting prominence of an extracellular matrix and secretion of laminin. Antoni B tissue is more loosely organized and most likely reflects degenerated Antoni A tissue.[4]

Diagnostic criteria include positive stains for S100 and SOX-10; staining for other classic markers such as CD34 and CD1117 must be negative. STAT6 immunohistochemistry, a reliable surrogate for detection of the NAB2-STAT6 fusion gene seen in solitary fibrous tumors, must also be negative. Negative markers for smooth muscle (Actin, Desmin) and negative melan-A will differentiate the tumor from the more common leiomyoma, and melanoma, respectively.[5]

Chemotherapy and radiation are ineffective for schwannomas and surgical enucleation is the only treatment. Video-assisted thoracoscopic surgical (VATS) excision is recommended for tumors less than 2 cm in size,[6] although one study used a VATS approach where the mean tumor size was 3 cm.[7,8] Larger tumors typically require esophagectomy with esophagogastrostomy. In this case, only a small portion of the tumor was adherent to the esophagus and primary excision and repair was performed.

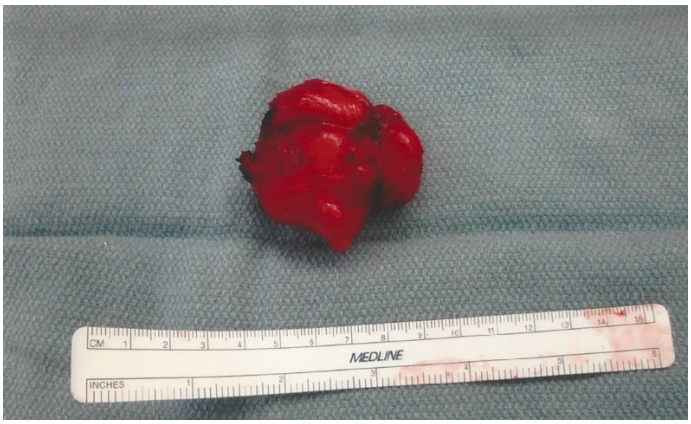

Figure 1. – Excised Tumor

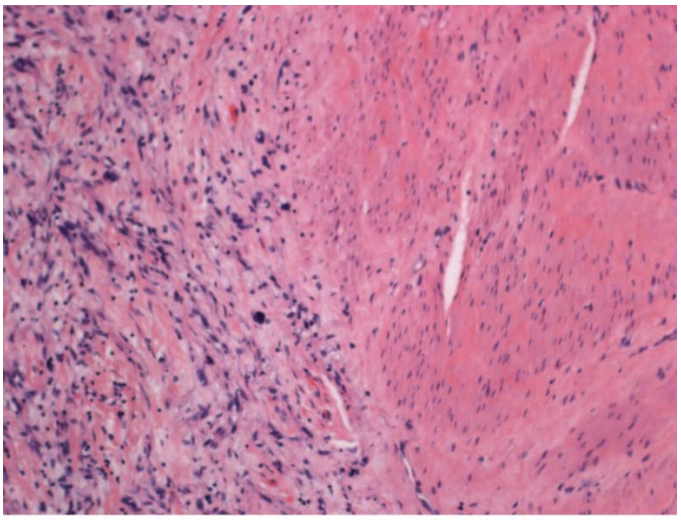
Figure 2. – Antoni A & Antoni B Tissues

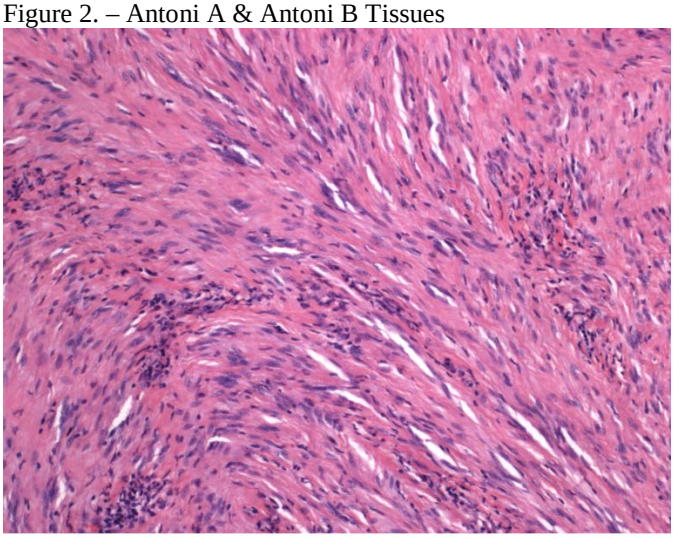
Figure 3. – Antoni A & Antoni B Tissues

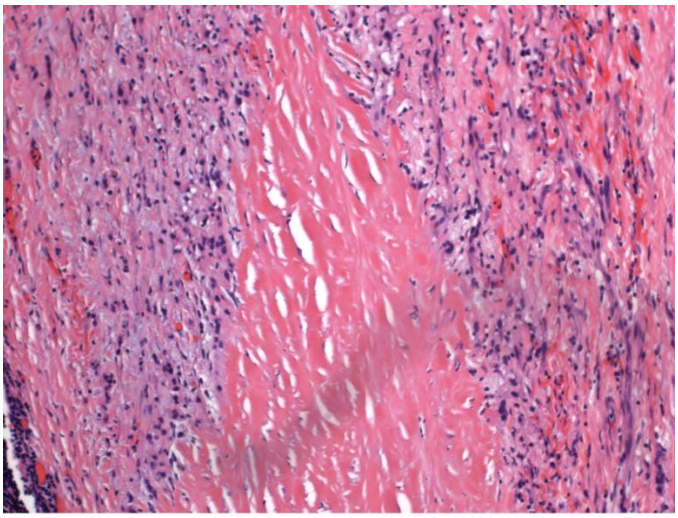
Figure 4. – Verocay Body

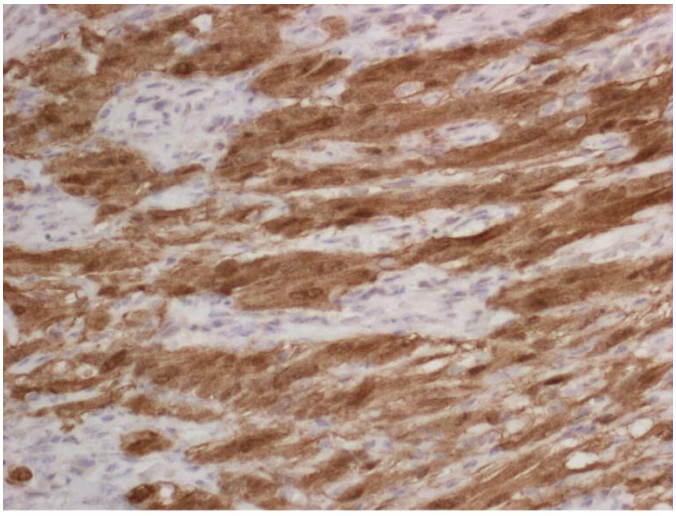
Figure 5. – S100 Staining

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