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Peripheral Nerve Sheath Tumor of the Esophagus. Case Report (1679)

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ABSTRACT

Peripheral nerve sheath tumors of the esophagus originate from the peripheral nerves in the esophageal plexus. They are extremely rare, slow growing, and cause progressive symptoms; schwannomas are the most common form. Histological studies and immunohistochemistry are required to diagnose and differentiate them from other tumors, including gastrointestinal stromal tumors, and as they are resistant to chemotherapy and radiotherapy, surgical resection is the treatment of choice.

We present a case of a man presenting with asymptomatic tumor in the posterior mediastinum that was discovered incidentally on a chest X-ray. The tumor was resected *en bloc* (esophagectomy), and as malignancy was not confirmed, reconstruction was deferred to a later date (gastric pull-up). The pathology analysis eventually confirmed it was a benign schwannoma that was immunohistochemically positive for S-100 and Ki-67. (J CANCEROL. 2016;3:28-31)

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INTRODUCTION

Tumors of the peripheral nerve sheath were first described in 1989 when Hirose, et al. identified a perineural cell tumor[1]. These types of tumors are composed of one or more of the cellular elements of the peripheral nerve, most commonly schwannomas and neurofibromas, the former consisting solely of Schwann cells and the latter containing fibroblasts, axons, and perineural cells in addition to Schwann cells[2]. Benign tumors are extremely rare in the esophagus, accounting for less than 2% of all neoplasms located in that site, and are mainly found in the middle third and the majority are leiomyomas.

CLINICAL CASE

A 34 year old man with no significant past medical history. During preoperative evaluation for aesthetic septorhinoplasty, he was incidentally found to have a tumor lesion in the mediastinum on a chest x-ray (Fig. 1). A computed tomography scan was done, which showed an 8 x 5 cm tumor in the posterior mediastinum, causing anterior displacement of the trachea, extending to the carina, and causing loss of interface with the esophageal wall (Fig. 2). A panendoscopy was performed that showed extrinsic compression 15 cm from the dental arch extending up to 3 cm above the gastroesophageal junction. An endoscopic ultrasound revealed tumor infiltration in the esophageal wall, with three 1 cm perilesional lymph nodes with metastatic appearance and 8 mm para-esophageal and celiac axis lymphadenopathies. An ultrasound-guided biopsy of the lesion was performed, which showed a tumor of the peripheral nerve sheath with uncertain malignant potential; immunohistochemistry was negative for CD-56 and positive for S-100 and Ki-67. On the basis of this evidence, surgery was decided and partial esophagectomy was performed with *en bloc* resection of the tumor, using a right posterolateral thoracotomy approach including gastrostomy and cervical esophagostomy.

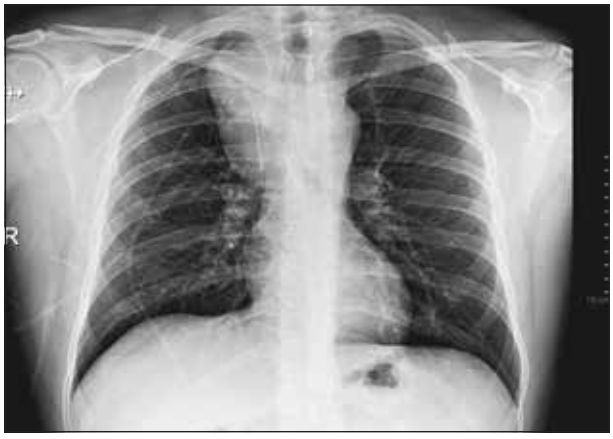

Figure 1. Chest X-ray on which mediastinal mass was seen incidentally.

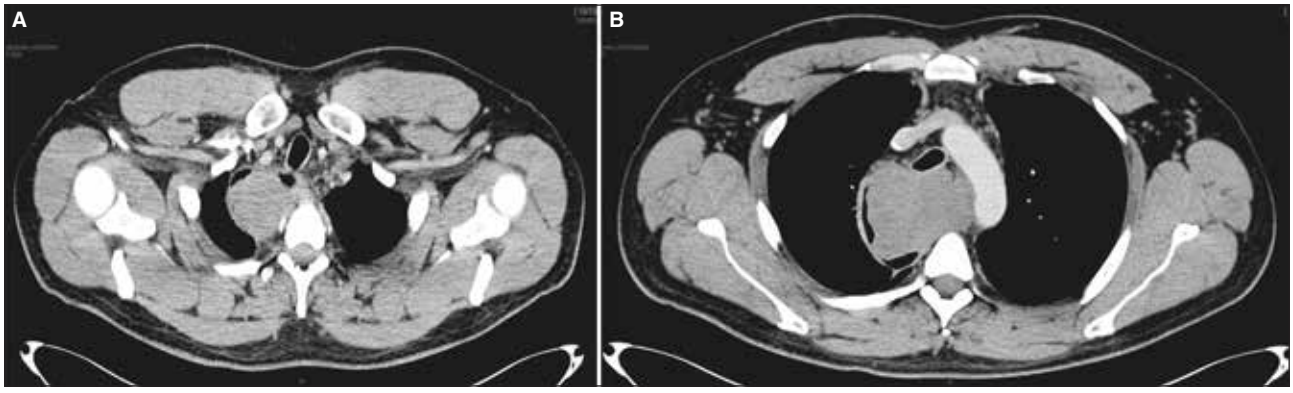

Figure 2. Chest CT scan showing a mass in the posterior mediastinum, with displacement of the trachea, extending to the carina and causing loss of interface with the esophagus.

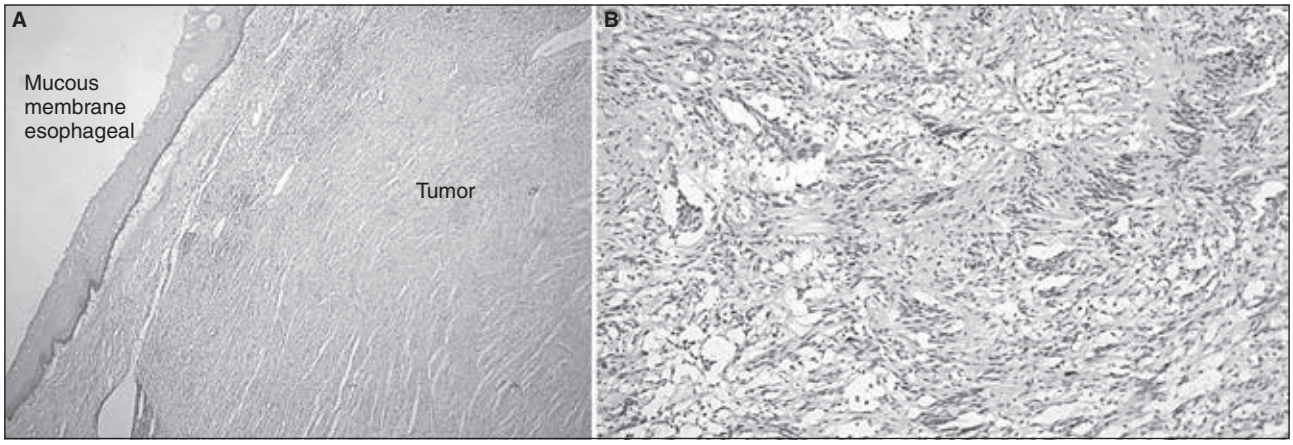

Figure 3. Histopathological images that confirm it is a peripheral nerve tumor with spindle shaped cells arranged in a palisade pattern. (H&E staining, 10x and 40x).

The findings were posterior mediastinum tumor measuring 20 x 16 x 10 cm, with infiltration of the right lateral esophageal wall, azygos vein, and ipsilateral vagus nerve. The pathology report confirmed it was a 10 x 8.5 x 5 cm benign peripheral nerve sheath tumor with degenerative changes, involving the esophageal wall, with negative surgical margins and with four lymph nodes negative for malignancy (Fig. 3). One month later, esophagogastric transit was restored through gastric pull-up with cervical anastomosis without complications. At present the patient is in follow-up and is asymptomatic.

DISCUSSION

The majority of tumors of the esophagus are malignant, so benign lesions are very rare; fewer than 1% are detected clinically and schwannomas, which were first described in 1967 by Chatelin, et al.[3], are extremely rare. They are slow-growing tumors with good prognosis, usually occurring between the third and seventh decades of life, with a similar distribution by gender, and originating from the esophageal nerve plexus. Preoperative diagnosis is difficult, so a definitive diagnosis cannot be made until after surgical resection. Patients present with dysphagia, dyspnea, and chest pain, which worsen as the tumor grows. In our case, in spite of the size of the lesion, the patient was asymptomatic and the tumor was found incidentally when a chest X-ray was taken. Imaging studies are very useful for confirming and for the delineation of the lesion in the mediastinum; however, determining whether the lesion is a schwannoma is difficult. In light of this situation, endoscopic ultrasonography has gained ground as it allows a biopsy to be taken of the tumor and diagnosis and treatment to be provided[4]. Histologically, schwannomas show one or two types of patterns, called Antoni A and Antoni B. The Antoni A pattern shows compact areas of spindle-shaped cells in palisade pattern, while the Antoni B pattern shows a loose arrangement of cells with cystic changes and hemorrhage. Immunohistochemically, schwannomas show positivity for S-100, CD-34, CD-117, and αSMA[5,6].

With regard to treatment, neither radiotherapy nor chemotherapy is effective, so the only option is surgical resection, including the capsule of the lesion. A minimally invasive approach can be used for lesions smaller than 2 cm, and thoracoscopic enucleation of the lesion is a procedure that provides the benefits of less postoperative pain and faster recovery. For larger lesions (greater than 8 cm), however, a thoracotomy approach with *en bloc*

resection is preferred, so frequently either an esophagectomy or esophagogastrectomy is performed, depending on the extent of the lesion[7,8]. In our case, the lesion measured 8 x 5 cm, so it was initially decided to perform a thoracotomy as it was not possible to determine if the tumor was benign or malignant. *En bloc* resection of the lesion was successfully achieved, and after ruling out malignancy, the patient underwent a second procedure for gastric pull-up reconstruction. Lastly, it is important to distinguish schwannomas from gastrointestinal stromal tumors as the latter have a high malignancy potential, the therapeutic strategy is different, and prognosis depends on the size and number of mitoses per high power field, as opposed to schwannomas, which have an excellent prognosis and only in very rare cases have there been reports of malignancy and metastatic nodules, based on histologic criteria that included a combination of factors such as mitotic activity, cellularity, nuclear atypia, and tumor necrosis[9,10].

CONCLUSION

Benign peripheral nerve sheath tumors of the esophagus are very uncommon, so even if suspected, other malignant entities must be ruled out and probably the only way to achieve this is by conducting a definitive histopathological study. Surgical resection is the treatment of choice and requires adequate preoperative planning in order to select the best approach and achieve complete resection in order to maximize tumor control.

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