Sympathetic chain Schwannoma

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ABSTRACT

الشوانوما (ورم غمد الليف العصبي)، هو ورم نادر، حميد، وبطئ في النمو. وينشئ هذا الورم من خلايا شوان (خلايا غمد الليف العصبي) التي تبطن غلاف الأعصاب. ندرج في هذا التقرير حالة مريض يبلغ من العمر 70 عاماً، يعاني من كتلة في العنق بدون أعراض أخرى. أسفر الفحص السريري عن وجود متلازمة هورنر في الجهة اليسرى وكتلة نابضة مع إزاحة أمامية للشريان السباتي الرئيسي. أظهر التصوير الطبقي المحوري (CT) وجود كتلة محددة الابعاد وبدون أخذ صبغة مع إزاحة وعائية. وأثناء الجراحة تبين أن الورم كان ناشئاً من الضفيرة السمبثاوية، وقد تم استئصالها بشكل كامل. أكد الفحص النسيجي تشخيص الشوانوما أنطوني (أ) وأنطوني (ب). وكان المريض في حاله جيده و خال من أية انتكاسات للورم لمدة 14 شهراً. نستعرض في هذه الورقة التقييم السريري، الإشعاعي، والنسيجي، إضافة إلى العلاج والمضاعفات الجراحية لهذا الورم.

Schwannomas are rare, benign, slowly growing tumors arising from Schwann cells that line nerve sheaths. Schwannomas arising from the cervical sympathetic chain are extremely rare. Here, we report a case of a 70-year-old man who presented with only an asymptomatic neck mass. Physical examination revealed a left sided Horner syndrome and a neck mass with transmitted pulsation and anterior displacement of the common carotid artery. Computed tomography (CT) showed a well-defined non-enhancing mass with vascular displacement. The nerve of origin of this encapsulated tumor was the sympathetic chain. The tumor was excised completely intact. The pathologic diagnosis was Schwannoma (Antoni type A and Antoni type B). The patient has been well and free of tumor recurrence for 14 months with persistence of asymptomatic left sided Horner syndrome. The clinical, radiological and pathological evaluations, therapy and postoperative complications of this tumor are discussed.

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Schwannoma (Neurilemmoma) is a rare neurogenic neoplasm originating from nerve sheaths and can arise from any myelinated nerve with rare malignant degeneration.[1,2] Approximately 25-45% of all Schwannomas occur in the head and neck.[3] Extracranial Schwannomas occurring in the head and neck region may arise from cranial, peripheral or autonomic nerves.[4] Determination of the nerve of origin is often difficult and not often made until the time of surgery.[4] Preoperative diagnosis may be difficult and needs a high index of clinical suspicion.[2] Extracranial Schwannomas usually present as solitary and well-demarcated lesion with insidious course. Schwannomas arising from the cervical sympathetic chain are extremely rare.[4] Computed tomography and MRI are the best diagnostic tools.[5] Surgical excision is the treatment of choice with low recurrence rate.[1,2,6]

Case Report. A 70-year-old man presented with a 10-year history of left sided neck swelling with no stigmata or family history of neurofibromatosis type 1 or 2. The size remained stationary over this long time with no associated pain, headache, dyspnea, dysphagia or hoarseness. Patient denied a history of trauma or other swellings. Systemic review was unremarkable. The patient was not diabetic, hypertensive or having episodes of transient ischemic attacks. On examination, there was a 9 cm oval swelling along the anterior border of the left sternomastoid muscle with transmitted pulsation (**Figure 1**). The left common carotid artery was displaced anteriorly. There was a crease scar of a previous incisional biopsy which was done elsewhere. There was no thrill or bruit, but left sided Horner syndrome was evident. Oral examination revealed no bulging. Systemic examination showed no stigmata of neurofibromatosis. Indirect laryngoscopy revealed mobile vocal cords. Basic blood investigations were within normal limits. Chest x-ray showed normal copula of diaphragm. Computed tomography revealed a 9 cm well – defined mass without contrast enhancement and with anterior displacement of the common carotid artery (CCA) and the internal jugular vein (IJV) (**Figure 2**). Revision of the previous biopsy confirmed a diagnosis of Schwannoma. Through a longitudinal incision along the anterior border of left

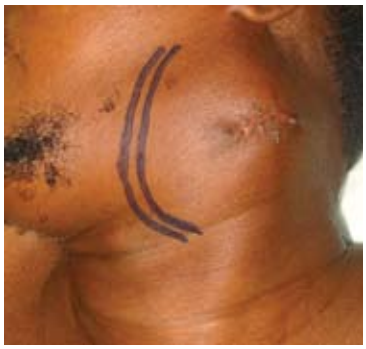

Figure 1 - An oval swelling along the anterior border of left sternomastoid muscle displacing common carotid artery anteriorly with scar of biopsy.

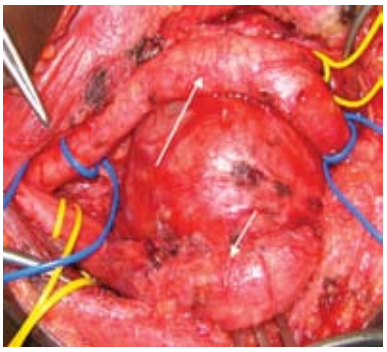

Figure 3 - Encapsulated Schwannoma displacing common carotid artery (long arow), and internal jugular vein (short arrow) anteriorly.

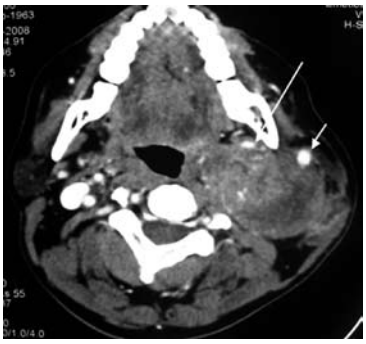

Figure 2 - Computed tomography scan showing a 9 cm mass without enhancement and displacing common carotid artery (long arrow) and internal jugular vein (short arrow) anteriorly.

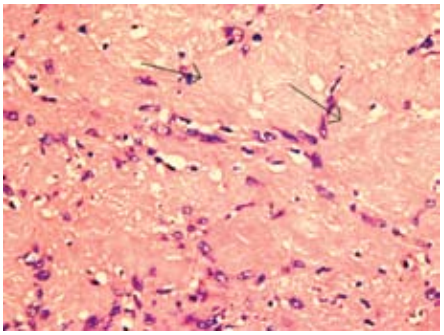

Figure 4 - Schwann cells arranged around an esinophilic material, verocay bodies (arrows) (Hematoxylin and Eosin, x 400).

sternomastoid muscle, a big oval, encapsulated mass was found displacing the common carotid artery, the IJV and the vagus nerve anteriorly (**Figure 3**). The nerve of origin was the cervical sympathetic chain. The vagus nerve, hypoglossal nerve, recurrent laryngeal nerve and superior laryngeal nerve were intact. The mass was dissected from the common carotid artery, IJV and vagus nerve and excised completely. Postoperatively, patient had an uneventful recovery, with persistent asymptomatic left sided Horner syndrome. Histopathological examination confirmed a diagnosis of Schwannoma, exhibiting features consistent with Antoni type A and Antoni type B (**Figure 4**). Postoperative Follow–up for 14 months showed no recurrence.

Discussion. Schwannomas are rare neurogenic tumors.[5,7] They originate from Schwann cells that line nerve sheaths and can arise from any myelinated nerve, therefore the olfactory and optic nerves are never affected.[1,3] They may arise from peripheral, cranial or autonomic nerves.[4] They are usually benign, solitary and run a slowly growing course.[1-3,5] Approximately 25-45% of all Schwannomas occur in the head and neck.[3] The parapharyngeal space, which is a well-defined anatomic zone of loose areolar tissue lateral to the pharynx, is the most common site of tumor origin.[6,8] The most frequently affected nerve is the eighth.[7] The nerve of origin could be the vagus nerve, sympathetic chain, hypoglossal nerve, ansa cervicalis, superior laryngeal nerve, glossopharyngeal nerve, facial nerve, deep cervical plexus or any other nerve in the cervical region.[5,9] Seventy seven percent of patients present with a unilateral cervical mass.[9] Pressure symptoms are the next most common mode of presentation, and are often a helpful indicator of the nerve of origin.[9] The mean delay between symptomatology and treatment is 5 years.[5] Preoperative diagnosis is usually difficult and in most cases is made at the time of surgery.[4,5] The nerve of origin is identified in 47-62% of patients who underwent surgery.[8,9] Schwannomas may mimic the physical and radiological findings of carotid body tumors.[4] Preoperative imaging modalities include ultrasound (US), CT and MRI. US is the least sensitive, while CT and MRI are the most relevant diagnostic tools.[5] Preoperative diagnosis may be aided

by fine-needle cytology, MRI or CT.[6] These tumors are usually encapsulated.[3] The treatment of choice of these tumors is complete surgical excision with the intent to preserve the nerve of origin.[1,2,6] Malone et al reported a complete excision in 88% of cases, while 12% had subtotal resection.[8] For extensive tumors involving the base of skull, middle ear or facial nerve, subtotal resection or nerve sacrifice with reconstruction and rehabilitation should be considered.[6,8] The most significant postoperative morbidity is associated with the Schwannomas of the vagus nerve, sympathetic chain, hypoglossal nerve, glossopharyngeal nerve and the facial nerve.[9] These tumors are resistant to radiotherapy.[1] Recurrence after surgical excision is rare, even after incomplete resection.[2]

In conclusion, cervical Schwannomas are rare neurogenic tumors, of neural crest origin, that need surgical intervention with rare recurrence after complete, or even incomplete, excision. Pre–operative CT and MRI are essential to formulate a precise anatomical mapping and to inform the patient about the potential hazards.

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