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Extracranial Schwannoma: An Unusual Case of Left Neck Mass

Thiviya. M[1], Dr Nurhamizah A[2]

[1,2]Otolaryngology Hospital Shah Alam, Malaysia

Abstract:
Schwannomas are benign neurogenic tumors that develops slowly from the myelin-producing Schwann cells of the peripheral sensory nervous system. Hypoglossal schwannoma is rare, accounting for about 5% of cases and typically with limited imaging characteristics. We report a unique case of a 41 years old female presented with asymptomatic swelling at left submandibular area. Magnetic resonance imaging revealed two well encapsulated neck masses in the left submandibular and left post-styloid parapharyngeal spaces, suggestive of Schwannoma, with unknown nerve origin. The patient underwent surgery, and a histological examination confirmed a schwannoma of the hypoglossal nerve. Compressive diagnostics is required for accurate diagnosis and of this rare clinical entity.

Keywords: Schwannomas, Hypoglossal Nerve, Submandibular

Case report:

Introduction

Schwannomas are slow growing encapsulated tumor that appear from cells that produce myelin sheath that covers peripheral nerves. There are 2 types of Schwann cells which are Antoni A and Antoni B. (1) It usually arises from patients between the age of 30 to 50 years old and equally affected on both sexes. Around 25-45% of schwannomas develop in the head and neck. (2). It's a rare tumor where by it represents about 5% of soft tissue tumors. The usual cranial nerve which gets affected are V, VII,X, XI and XII and sympathetic chain .(3) The most common cranial nerve that affected is vagus which is about 37% and the most least affected is accessory nerve which is about 4%.(4) Hypoglossal schwannoma has a higher female prevalence especially on middle aged patients .(5) Majority of schwannomas are asymptomatic but they may present with dysphagia if involving parapharyngeal space .Magnetic Resonance Imaging plays the most important role in diagnosing nerve neoplasm .Accurately determining tunor size, nature , and its antomical relationship with adjunct tissue and nerve courses is crucial for diagnosis and treatment planning .However this process can be hindered by rare entities that present with less specific radiological features. (6). The best treatment of preference is surgical excision of tumor and can be challenging for head neck surgeons as it can be troublesome in diagnosing precisely the condition before surgery and complication of surgical site. (5,7,8) Hereby we present a rare case of hypoglossal nerve schwonnama which was identified intraoperatively.

Case Report:

41 years old Malay female, no known medical illness, presented to Otorhinolaryngology Hospital Shah Alam with complain of left neck swelling for past five months in September 2022 with gradually

increasing in size. Patient had no complain of any pain, skin changes or skin ulceration. Patient has no dysphagia / odynophagia, no hoarseness of voice, nasal regurgitation, fever, loss of weight, loss of appetite, trauma and no history of familial malignancy. Examination reveals a soft, 6x4cm, left neck swelling at level 3 and 4, non-tender, with no skin changes. Oropharynx examination was remarkable and flexible scope shows normal vocal cord structures with no medialization. The very first fine needle aspiration was done and came out as left branchial cyst and subsequently was repeated with ultrasound guided biopsy and the result was nerve sheath tumour features favouring of schwannoma. Magnetic resonance imaging was done on October 2023 and reported as two well encapsulated neck masses in the left submandibular and left post-styloid parapharyngeal spaces as described and features are suggestive of Schwannoma, possibly of cervical sympathetic chain in origin and no obvious MRI evidence origin from glossopharyngeal, vagus or hypoglossal nerves. Then with the diagnosis, patient was operated under general and undergone excision of left neck schwannoma on December 2023 and noted it was hypoglossal origin. Post-operative mass measuring 10x4cm likely hypoglossal origin. Subsequently patient had no Horner syndrome but has cranial nerve hypoglossal palsy and requiring physiotherapy from speech therapist.

Discussion:

Generally, Schwannomas are rare peripheral nerve tumor, and it appeared at submandibular and post styloid space in this case. The submandibular space located at the superficial surface of mylohyoid muscle between the anterior and posterior bellies of digastric. The contents are submandibular gland, branches of facial and lingual artery, lymph nodes, and hypoglossal nerve. (6) It is important to know the exact location or anatomy of the tumor in order to preserve the function of the nerve. As mentioned earlier, the MRI reported as possibly of cervical sympathetic chain in origin and no obvious MRI evidence origin from glossopharyngeal, vagus or hypoglossal nerves but intraoperatively the tumor was attached with hypoglossal nerve. Hypoglossal schwannomas, tumors of purely motor nerves, comprise <5% of all head and neck schwannomas and quite common in middle aged patients especially women with mean age of 44 which is likely to our patient. Surgical management for this schwannoma can be graded based on imaging. The classification system for hypoglossal schwannoma includes 3 types which are type A (intradural type), type B (trans dural and extradural) dumbbell shaped tumors and type C for extracranial base tumors. In our case, the tumor was located at submandibular region and the tumor is categorised as type C schwannoma. (7)

As per clinical examination, it's a asymptomatic slow growing tumor that can be palpated at the left submandibular region in this case. Most patients diagnosed with schwannoma would not present with any neurological deficit as in this case and several differential diagnoses can be listed which are paraganglioma, branchial cleft cyst, malignant lymphoma, metastatic cervical lymphadenopathy. Since the patient was asymptomatic and had no any compressive symptoms, therefore the first diagnostic tool was done which is fine needle aspiration . The result came out as branchial cyst and repeated with ultrasound guided biopsy which reveals nerve sheath tumour features favouring of schwannoma. Nerve sheath is a layer of myelin and connective tissue that surrounds and insulates nerve fibers. A nerve sheath tumor is a growth within the cells of this covering. Nerve sheath tumors form in the layers of insulation which are schwann cells, endoneurium and perineurium. The nervous system made up of central nervous system and peripheral nervous system and nerve sheath tumors usually affects the peripheral nervous system. As in our case, the patient has schwannoma and in occurs in schwann cells and common locations

are head and neck. (8)

Due to rarity of extracranial hypoglossal nerve schwannomas and their similarity to neoplasm of submandibular salivary gland, paraganglioma, lymphoma and infectious disease a compressive diagnostic approach is essential. Distinguishing between these entities requires careful consideration of clinical findings and imaging studies for accurate diagnosis and appropriate management. (9) The diagnostic work up includes computed tomography (CT) and Magnetic Resonance Imaging scans (MRI) which were done in our case. In our case CT imaging was done at initial stage and subsequently proceeded with MRI and shows possibly of cervical sympathetic chain in origin in our case . The most effective imaging is MRI as it detects masses and assessing their extension as in our case. On MRI , T1 weighted images shows low intensity on schwannoma and T2- weighted images shows high signal. Usually, the target sign shows on T2 weighted images with increased peripheral signal intensity and decreased signal intensity. The increased signal intensity seen on T2- weighted images is typically caused by the presence of Antoni B fibres , known for their elevated water content relative to Antoni A fibres. (10) However surgical exploration is the best way to confirm diagnosis and total resection of the tumor was done in our case and it shows hypoglossal schwannoma Therefore it shows that surgical resection is always the best option and post operative , our patient had left tongue deviation due to the tumor .

Conclusion:

Hypoglossal schwannoma is rare tumor that have several diagnostic challenges and the treatment of choice is complete surgical excision with nerve sparing although this can be challenging as in our case . Prior to surgical procedure , complications of nerve involvements should be explained.

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