Pham et al., J Clin Exp Oncol 2013, 2:2
http://dx.doi.org/10.4172/2324-9110.1000107

Journal of Clinical & Experimental Oncology

Case Report A SCITECHNOL JOURNAL

Schwannoma of the Palatine Tonsil: A Rare Entity in an Eight Year Old Girl

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Abstract

Objective: We present a case of tonsillar schwannoma occurring in the youngest patient described in the English literature.

Results: An eight year old girl was found to have a 12 month history of slowly progressive dysphagia and two month history of a rapidly enlarging right sided oropharyngeal mass. A right sided tonsillectomy was performed and while there was an involved margin, the MRI post excision showed no residual tumour or abnormal enhancement. The final histology demonstrated the lesion to be a schwannoma.

Conclusion: This represents an important addition to the literature as there have only been eight other cases of tonsillar schwannoma described in the literature, two of which had been in children. This case represents the youngest patient reported to date.

Keywords

Schwannoma; Neurilemmoma; Palatine; Tonsil; Tonsillectomy

Case Report

An eight year old girl, otherwise well, presented with a 12 month of history of dysphagia (pharyngeal choking and gagging), and a two month history of an enlarging, pendulous right sided oropharyngeal mass. She was referred to the Department of Otorhinolaryngology, Head & Neck Surgery at Monash Medical Centre. A large, solid appearing mass was noted to be filling the oropharynx, appearing to be arising from the right inferior tonsillar/tongue base region. A connection to the left tonsil could not be excluded on examination whilst awake. Examination of the nose and nasopharynx were normal and there were no neck masses. The larynx could not be assessed due to the obstructing mass; however there was no history of hoarseness or breathing difficulty. A history of increasing snoring was also reported.

Pre-operatively, a differential diagnosis of lingual thyroid was considered and an ultrasound of the thyroid confirmed the presence of a thyroid gland in the neck. In addition, thyroid function tests were normal.

At operation, the mass was noted to be involving the inferior component of the right tonsil and a small portion of the right lingual tonsil (Figure 1). A capsular dissection of the right sided tonsil was performed with conventional monopolar diathermy encompassing the mass. The inferior component of the mass was removed separately and appeared to be arising from the region where the tonsillar branch of the glossopharyngeal nerve runs between the superior and middle constrictors. A very small nubbin was left at this region so as not to damage the nerve. The most likely diagnosis at the time was considered to be lymphoma; the left tonsil was not removed.

The specimen consisted of two parts, 5.5 x3 x2 cm and 2x1.5 x1cm, being the inferior component. Histopathology showed a lobulated uncircumscribed nodule covered in parts by a papillomatous epidermis. The lesion was composed of spindle cells arranged in hypercellular (Antoni A) and hypocellular (Antoni B) areas. There was peripheral palisading in amongst the spindle cells which were bland and wavy. There was no nuclear pleomorphism or mitotic figures. The histological appearance was in keeping with the diagnosis of Schwannoma (Figure 2). While it was incompletely excised, there was no evidence of lymphoma or malignancy. Flow cytometry and surface light chain analysis was inconclusive due to the paucity of B-cells.

Post-operatively, the patient made an uneventful recovery and was discharged one day postop. At the two month follow up, a MRI was performed and showed no mass lesion or abnormal enhancement in the right oropharynx and no evidence of schwannomas elsewhere. The patient was clinically free of disease with normal speech and swallow at five months follow up.

Discussion

Schwannoma or neurilemmoma are usually slow growing neoplasms which originate from the Schwann cells which myelinate nerve fibres. Between 25 and 48% have been reported to occur in

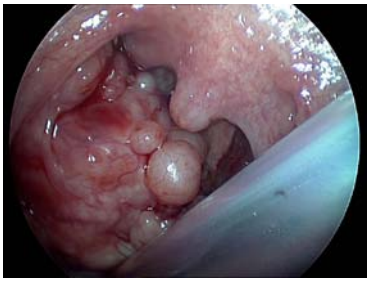

Figure 1: Right oropharyngeal mass causing dysphagia.

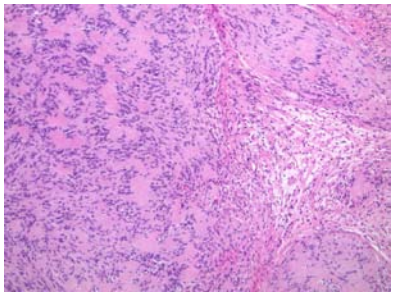

Figure 2: HypercellularAntoni A and hypocellularAntoni B areas.

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Received: December 18, 2012 **Accepted:** January 19, 2013 **Published:** January 26, 2013

the head and neck [1,2] although they can originate from any site which contains a neural sheath. In the head and neck it occurs most commonly with the acoustic nerve and is rarely found in the oral cavity or oropharynx [3]. Schwannoma of the tonsil is rare, with only eight cases being reported previously in the English literature [3-10]. Malignant transformation of a schwannoma is rare [11].

In the head and neck the most commonly involved nerve is the acoustic nerve. Others in descending order are the trigeminal, facial and hypoglossal nerves. They do not arise from the olfactory or optic nerves because these nerves lack Schwann cells. Tonsillar schwannomas are presumed to arise from a branch of the glossopharyngeal nerve [10] and are all unilateral [3-10].

There is no recognised risk factor for this condition and the ages of presentation can range from eight to 69 [6].

Symptoms are that of slowly progressive dysphagia, which seldom produces obstruction to respiration although obstructive sleep apnoea has been reported [8]. Symptoms, however, can be as short as three months for dysphagia [5]. One patient noticed growth over four years [6]. Bleeding is uncommon and paraesthesia along the distribution of the nerve, foreign body feeling in throat, odynophagia and snoring have also been described [3,8].

Macroscopically the schwannoma of the palatine tonsil can reach diameters of 5 cm [3] or 5.5 cm with this present case. Histologically, schwannomas are composed of alternating Antoni A and Antoni B areas. Antoni A areas are composed of spindle shaped cells in rows, with nuclear palisading or Verocay bodies. Antoni B cells and fibres are arranged in a disorderly pattern, with areas of microcysts and fluid accumulation [7]. Vascularity is not a prominent feature and necrosis and mitotic activity are seldom seen [8]. Tumour cells are strongly and diffusely immunoreactive for S-100 protein [2].

CT has been used as first line to assess tonsillar schwannomas. All describe a well circumscribed mass, with heterogeneity [7,8,10] and hypovascularity [3]. MRI however is the gold standard of diagnosis for head and neck schwannoma with 100% accuracy [2,12].

The treatment for tonsillar schwannoma is surgical excision, although wide excision is not necessary due to the benign nature of the lesion. Schwannomas are radioresistant therefore radiotherapy plays no role in treatment [13].

Compared to the eight known cases of schwannoma of the tonsil, this case presents the youngest case of schwannoma of the tonsil in the literature [3-10]. Interestingly, this case also presents a tonsillar schwannoma with the known widest diameter of 5.5 cm. There is a slight right sided predilection with the other cases and this case is also right sided. All cases were similar in that they were unilateral, were treated with surgical excision alone and had no recurrence.

Conclusion

We present a case of schwannoma of the palatine tonsil in an eight year old girl, the youngest patient reported to date.Tonsillar schwannoma is a rare occurrence, and may be included in the differential diagnosis of a unilateral tonsillar mass. The gold standard of detection is with MRI and the diagnosis is confirmed with microscopy. The gold standard of treatment is with surgical excision. This case report represents the youngest patient in the literature with the diagnosis of tonsillar schwannoma and represents an important addition to the literature on this rare condition.

Summary

- Schwannomas are benign tumours that arise from Schwann cells that myelinate nerve fibres.
- They commonly occur in the head and neck, and are mostly associated with the acoustic nerve.
- Schwannoma of the palatine tonsil is rare, with only eight cases previously reported in the English literature.
- Diagnosis is by histopathology, and the gold standard of treatment is with surgical excision.
- We present a case of schwannoma of the palatine tonsil in an eight year old girl, the youngest patient reported to date.

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