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Case Report

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Pediatric Thyroid Schwannoma: A Case Report

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Abstract

A case of a thyroid schwannoma in a 12-year-old male is described. An asymptomatic right sided neck mass was initially seen on telemedicine as a shadow. After a workup; the patient underwent an en bloc excision of the mass, pathology was consistent with a benign schwannoma. To our knowledge this is the 5th case of a thyroid schwannoma in a pediatric patient described in the literature since the first case was reported in 2004.

Introduction

Shwannomas are the most common neurogenic tumor, originating from nerve sheath cells. They are benign, slow growing neoplasms that can occur anywhere in the body that contain from nerve roots. Extracranially, 25-40% are found in the head and neck. Peak incidence is in the third to fifth decade of life, men and women are affected equally [1]. Thyroid involvement presents an infrequent circumstance, even rarer in the pediatric population. The first case of a pediatric thyroid schwannoma was described in the literature in 2004 [2], since then there have been 3 other cases [3-5]. Here we present a 12-year-old male with an asymptomatic thyroid schwannoma.

Case Presentation

A 12-year-old male presented to the office for evaluation of right neck mass initially noted as a 'shadow' on a telemedicine visit one month prior. Mother reported that since discovery there have been no associated symptoms including dysphagia, dysphonia, or hoarseness. The mass was clinically stable in size. The patient denied skin changes, drainage, erythema, pain, or any other associated symptoms. There was no family history of thyroid disease or soft tissue masses. The patient was otherwise in good health. Physical exam revealed a right-sided fixed, nontender mass, approximately 3 × 2 cm, with no associated cervical lymphadenopathy. It did not move with swallowing. Ultrasound revealed a 16 mm nodule arising from, or bordering, the right thyroid lobe. Labs were unremarkable. Subsequently an MRI of the neck with and without contrast was obtained to better characterize the mass and its anatomic relations. This revealed a soft tissue mass anterior to the border of the right thyroid lobe, that did not appear to be contained within the thyroid itself. The mass demonstrated homogeneity, mildly enhancing with contrast. The left side of the thyroid was unremarkable (Figure 1 and Figure 2). A fine-needle aspiration of the mass was performed that identified spindle cells and prominent metachromatic fibrillary stroma suggestive of Verocay bodies. The specimen stained positive for S100. Pathology was consistent with spindle cell neoplasm - the differential diagnosis included a schwannoma, neurofibroma, and other spindle cell neoplasms.

The patient underwent a neck exploration that revealed a densely adherent mass involving the capsule of the right thyroid lobe, with no evidence of invasion into adjacent soft tissue or crossing the midline. The mass was resected en bloc with a portion of the thyroid gland. There was an area of muscle inferiorly that the mass was densely adherent to that was resected separately to ensure a negative margin. The specimen was sent for frozen intraoperative pathology which was consistent with spindle cell neoplasm favoring schwannoma. After discussion with pathology and our feeling that adequate margins were achieved, we did not believe a thyroid lobectomy was warranted. The final pathology showed spindle cell proliferation with predominant Verocay bodies, strong positive stain for S100 - consistent with a benign schwannoma. All margins were free of tumor. The patient remains asymptomatic with no evidence of recurrence at this time.

Case Report

A schwannoma is a tumor that arises from Schwann cells of the peripheral nerves, most commonly the sensory nerves. The most frequent schwannoma of the head and

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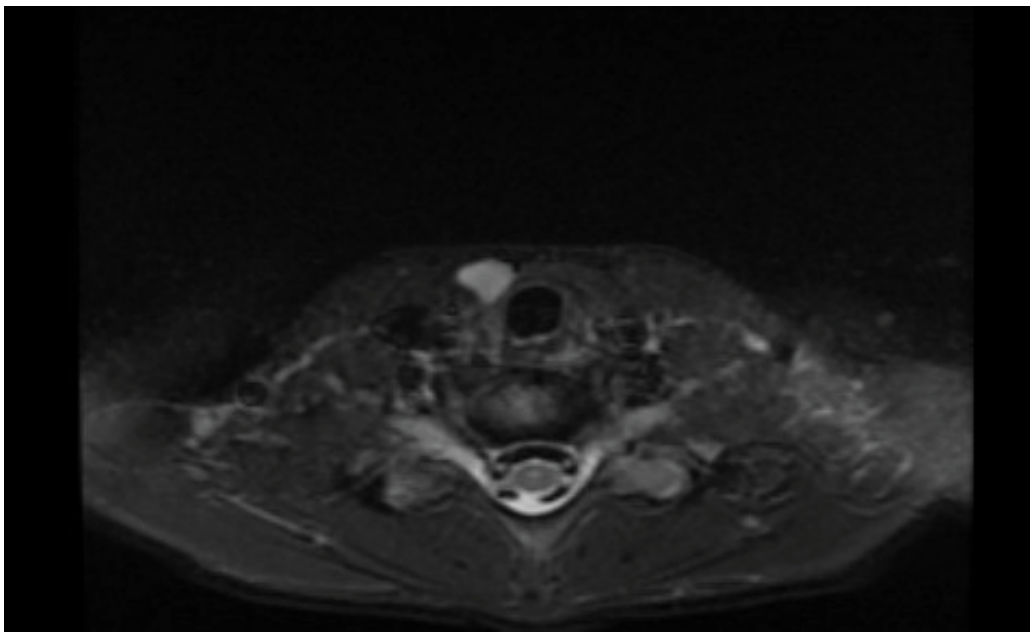

Figure 1: Axial T2 weighted image showing high signal right sided nodule immediately anterior to thyroid

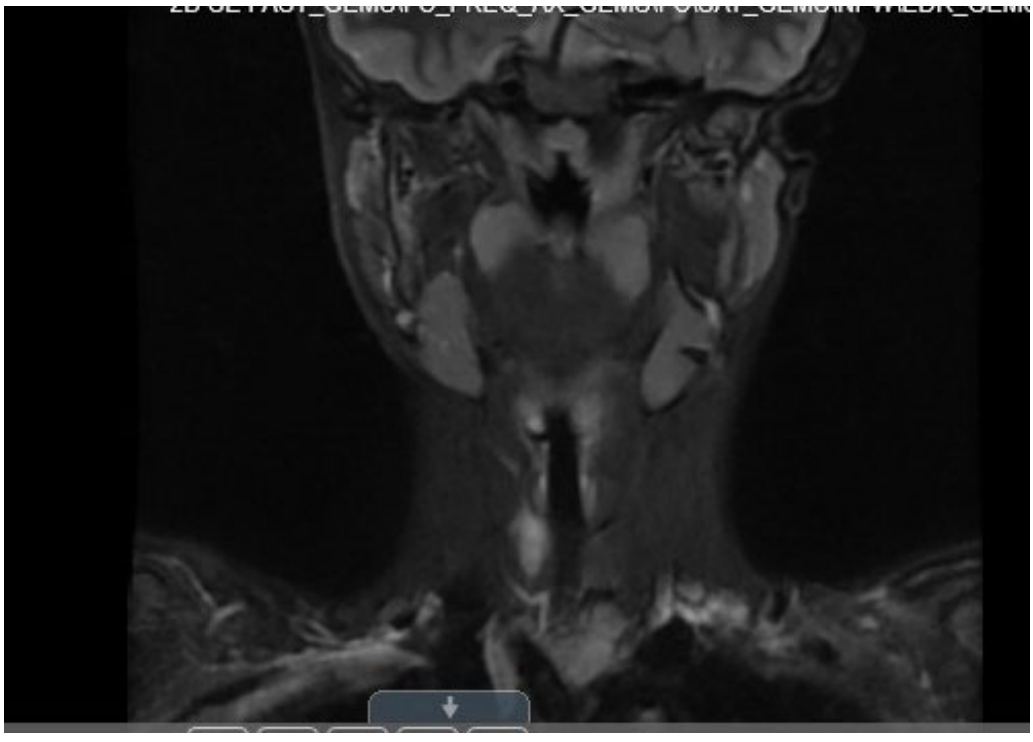

Figure 2: Coronal T2 weighted image of right thyroid nodule.

neck is a vestibular schwannoma, most frequently arising from the superior vestibular nerve [6]. However, only 25% of schwannomas occur within the head and neck region, and are even rarer within the thyroid gland [7]. They are further divided based on their histologic features into Antonio A and Antonio B categories. Antonio A tumors consist of palisading nuclei around ventral cytoplasm, and Antonio B tumors are comprised of a loose edematous matrix[8] and typically present in the parapharyngeal space.

Workup of these tumors often includes both CT scan and MRI. CT scan allows visualization of the association of the mass with the boney architecture and the presence of calcification. MRI enables soft tissue visualization to better assess the relationship with neurovascular structures. Large schwannomas may require angiography to locate feeding vessels that potentially require preoperative embolization [8]. However, many patients initially presenting with a thyroid nodule undergo a thyroid ultrasound. These tumors

can be difficult to distinguish from a thyroid nodule using this imaging study, and therefore are commonly diagnosed after surgical intervention.

The treatment of a schwannoma is enucleation of the tumor. It is important to attempt to preserve nerve function during removal, however this can be difficult if the tumor is large. The risk of malignancy is 4%, and therefore the potential morbidity of enucleation must be taken into consideration against the risk of tumor growth [8].

There have been 19 reported cases of thyroid schwannomas. The first was reported by Delaney and Fry in 1964. More recently, Dhar, et al. discussed the case of a thyroid schwannoma that presented as a benign thyroid nodule. Histology showed Antoni A (hypercellular areas) with Verocay bodies and Antoni B (hypocellular pattern). The lesion arose from within the thyroid gland itself and not extrinsically. Primary thyroid schwannomas are the rarest subset of neck schwannomas. They are believed to arise from intrathyroid sensory nerves or from the autonomic innervation to the thyroid [5]. They are often mistaken for thyroid nodules because they both share clinical and sonographic characteristics.

Thyroid gland schwannomas are quite uncommon. These are seen even more infrequently within the pediatric population. The first case of thyroid schwannoma in a pediatric patient was reported in 2004 in a 12-year-old girl [3]. This was followed by a case report concerning a 14-year-old male in 2010 [4]. The last reported case occurred in 2017 in a 12-year-old female that presented with neck swelling after upper respiratory infection [3]. In one of these cases, fine needle aspiration was performed on two occasions with the cytology classified as Bethesda Category 4, “suspicious for Hurthle cell follicular neoplasm”. Ultrasound images were concerning for malignancy, leading to surgical intervention. She subsequently underwent a right thyroid lobectomy. Final pathology revealed a thyroid schwannoma, as the tumor stained positive for S-100 and was negative for TTF-1 [3].

Conclusion

Thyroid schwannomas are an exceedingly rare head and neck tumor, with even fewer cases reported in the pediatric population. They frequently present as a painless neck mass and can be difficult to distinguish from thyroid nodules on ultrasound because they share clinical and sonographic characteristics. As such, the diagnosis is often made post-operatively on surgical pathology. It is important to consider a thyroid schwannoma among the differential diagnoses when evaluating pediatric thyroid nodules in the clinic.

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