

Plexiform schwannoma embedded within the masseter muscle: A case report

Soroush Farnoosh, BS^{a*}; Debra Don, MD^b; Masoud Shamszadeh, MD^c; Jeffrey Koempel, MD^b

***Soroush Farnoosh, BS**

University at Buffalo School of Medicine and Biomedical Sciences. Buffalo, NY, USA

Phone: +1-323-361-2145; Email: soroushf@buffalo.edu

Abstract

Schwannomas are benign nerve sheath tumors. Up to 45% of the schwannomas occur in the head and neck and 5% of schwannomas will develop into a rare form called plexiform schwannoma. We managed a 10-year-old patient with a plexiform schwannoma embedded within her masseter muscle by surgically removing the mass without any adverse sequelae. Plexiform schwannomas should be considered in the differential diagnosis of mid-cheek masses. A proper diagnosis preoperatively would ensure that conservative measures are taken to cause minimal damage to the nerves or be prepared to do nerve grafting if harming of the nerves is inevitable.

Keywords

Schwannoma; Plexiform schwannoma; Nerve sheath tumor

Background

Schwannomas are rare, benign, and encapsulated nerve sheath tumors that arise from myelin producing Schwann cells. Studies have reported that 25-45% of all schwannomas occur in the head and neck region [1][2]. The plexiform schwannoma is a rare subtype of schwannoma first described by Harkin et. al. in 1978 [3]. It is estimated that approximately 5% of schwannomas may further evolve into the plexiform schwannoma [4]. The plexiform schwannoma owes its name to its multinodular appearance both microscopically and macroscopically. They occur more commonly in children and young adults and can be seen in the setting of neurofibromatosis-2 and schwannomatosis. The plexiform schwannoma has a predilection for the head and neck and although usually subcutaneous, can also occur at mucosal sites including lips, tongue and hard palate. Here we presented a rare case of plexiform schwannoma embedded within the masseter muscle followed by a brief review of the literature.

History

A 10-year-old child came to the ENT department with a two-year history of a slowly enlarging mass in her left cheek. She did not have any associated symptoms and on exam, she was noted to have a

2 x 3 cm firm, well-circumscribed mass in the just above the body of the mandible. The mass was not tender and had no overlying skin changes. Her left facial nerve had normal function.

Radiological Findings

A CT scan of the neck revealed diffuse enlargement of the left masseter muscle with mild heterogeneity and some enhancement. An MRI of the head and neck also showed a lobulated mass involving the left masseter muscle with marked enhancement of the mass on T2 weighted images. Nodular organization was noted in T1- and T2-weighted images with fat suppression. T2-weighted images with fat suppression also showed a heterogeneous nodular formation with a hypointense rim relative to the nodular core.

Surgical Procedure

The mass was approached via an extended parotidectomy incision wherein extensions were performed into the hair-bearing area of the scalp and the cervical region. The tumor was visualized after subplatysmal flaps were raised. Stenson's duct, as well as several buccal branches of the facial nerve, were found to be coursing over the lateral aspect of mass. The mass appeared anterior and distinct from the parotid gland. Facial nerve monitoring was used to identify and preserve the facial nerve branches. The mass was well-encapsulated and separated easily from its surrounding structures.

Pathological Finding

The tumor consisted of a lobulated fragment of tan-grey firm tissue measuring 4.3 x 3.5 x 2.5 cm. Microscopic evaluation with H and E stain revealed a cyst with distinct encapsulated nodular organization. In addition Antoni A regions and Verocay bodies were identified in the specimen. Immunohistochemical staining of the specimen revealed positive and diffuse staining for CD56 and calretinin, but sparse and patchy staining for Factor XIIIa and CD34. (Image 2)

Discussion

The differential diagnosis of mid-cheek masses includes both benign and malignant lesions that can originate from any of the soft tissues of the face – including adnexal, lymphatic, neurogenic and salivary structures. The most common neoplasm encountered in the mid cheek is the accessory parotid gland tumor. Neuromas, although not very common, should always be included in the differential diagnosis because of their close associations with nerves in the head and neck region and possibility of damage to those nerves during surgical interventions. More common than the neuromas with neuronal origin, are neuromas arising from the tissues accompanying the neurons. The term “peripheral nerve sheath tumor” then basically describes pathologically unique tumors including benign lesions such as neurofibroma, schwannoma, perineurioma, traumatic neuroma, or malignant peripheral nerve sheath tumor (MPNST). Among these, schwannomas and neurofibromas are the commonest benign forms. Schwannomas, as the name indicates, are caused by hyperplasia of the myelin producing cells in the peripheral nervous system, or also know as Schwann cells, named after a German histologist Theodor Schwann. In contrast, neurofibromas contain more than just the Schwann cell and may involve axons, perineural cells, or fibroblasts.

Macroscopically schwannomas are round and encapsulated tumors presenting with tan colored and homogeneous looking areas. They can also present as cystic or hemorrhagic but with no gross

necrosis. Schwannomas have been categorized into conventional, cellular, plexiform, and melanotic. Plexiform schwannoma can refer either to conventional or cellular schwannoma and simply indicates the nodular organization of the tumor, which arises from multifocal growth of Schwann cells within the nerve fascicles. Plexiform schwannoma was shown to represent 4.3% of all schwannomas, and 23% of the head and neck schwannomas [5].

All types of schwannomas share certain microscopic characteristics. Nils Rangnar Eugene Antoni was the first to describe the microscopic characteristics of some peripheral nerve sheath fibers, which later became known as schwannoma. Antoni described 2 forms of cellular arrangement, named after him Antoni A and Antoni B regions, which have become pathognomonic of schwannoma. Antoni A regions include hypercellular areas with cells looking like spindles and form tightly organized rows named nuclear palisade. Two parallel nuclear palisades with an anuclear zone in the middle form a Verocay body. In contrast, Antoni B regions include microcytic, loosely packed hypocellular areas [6]. Although microscopically differentiable, there may be some morphological overlaps between plexiform schwannomas and plexiform neurofibromas. Thus immunohistochemical markers have been developed to diagnose and distinguish the two, including markers for S100, CD34, calretinin, factor XIIIa, and CD56. Although S100 is present in both neurofibroma and schwannoma, factor XIIIa and CD34 are more exclusive for neurofibromas, whereas calretinin and CD56 are more indicative of schwannoma [7][8].

Radiological presentation of schwannomas is well known and has been described vastly in the literature. In a study done by Lee et. al. [9] all the schwannoma cases studied demonstrated well circumscribed low attenuation on CT scans, but this presentation is shared among many other benign tumors as well. But magnetic resonance imaging, especially with gadolinium contrast and fat suppression, has been shown to be of tremendous diagnostic value [10][11]. Schwannomas have been described as having isointense signal intensity on T1-weighted images, but hyperintense signal intensity on T2-weighted images. A characteristic target shape has also been described in some cases of schwannoma on T2-weighted or gadolinium-enhanced T1-weighted images [9][12]. Also intralesional hypointensities between the nodules are suggestive of fibrous septae between them [8]. Although radiologists may commonly consider including schwannoma in the differential diagnosis, determining the plexiform characteristics of schwannoma might be somewhat challenging. Even so, Hebert-Blouinet. al. [11] claim that careful inspection of the MR images can unveil the structural quality of plexiform schwannomas. They further report that plexiform schwannomas present with enlarged fascicles coursing through and beyond the primary lesion, with a more tubular appearance, and nodules collected around a single nerve or fascicle. They also reported a few cases of plexiform schwannomas with the characteristic target shape.

The first line of treatment for benign neural sheath tumors is surgical excision of the mass, but priority is given to preservation of accompanying nerves. And since malignant transformation is rare, radical excision of the surrounding area is not indicated. Although plexiform schwannomas are not malignant, recurrence may happen if not completely excised [13]. Ideal surgical incisions should provide adequate exposure, while preserving the aesthetic features of the face. Superficial mid-cheek masses commonly encircle the branches of the facial nerve and/or Stensen's duct, thus adequate exposure is critical for surgical management of the mid-cheek masses. For the same reason, intraoral incisions are avoided for masses superficial to the buccinator muscles. The common recommendation is

an extended parotid-submandibular incision which provides adequate exposure for extended areas of the face, while preserves cosmetic features and has little to no complications [14]. Many surgical techniques have been described for excision of head and neck schwannomas including: Complete tumor excision and the accompanying nerve(s) followed nerve grafting, tumor excision with sparing the nerves; tumor enucleation between adjacent nerves; tumor emptying and saving the capsule, “shelling out” of the tumor, and nerve-sparing subcapsular resection technique [15]. Since none of these techniques are flawless and postoperative complications may occur with any of them, the surgeon should make the final decision based on the anatomic position of the tumor after meticulous observation of the nerves passing adjacent to the mass.

Conclusion

In conclusion, schwannomas should always be considered in the differential diagnosis of mid-cheek masses and since they always arise near neural tissue, a proper diagnosis preoperatively would ensure that conservative measures are taken to cause minimal damage to the nerves or be prepared to do nerve grafting if harming of the nerves is inevitable. This case represents a great example for having both the pathological and radiological presentations of plexiform schwannomas.

Figures

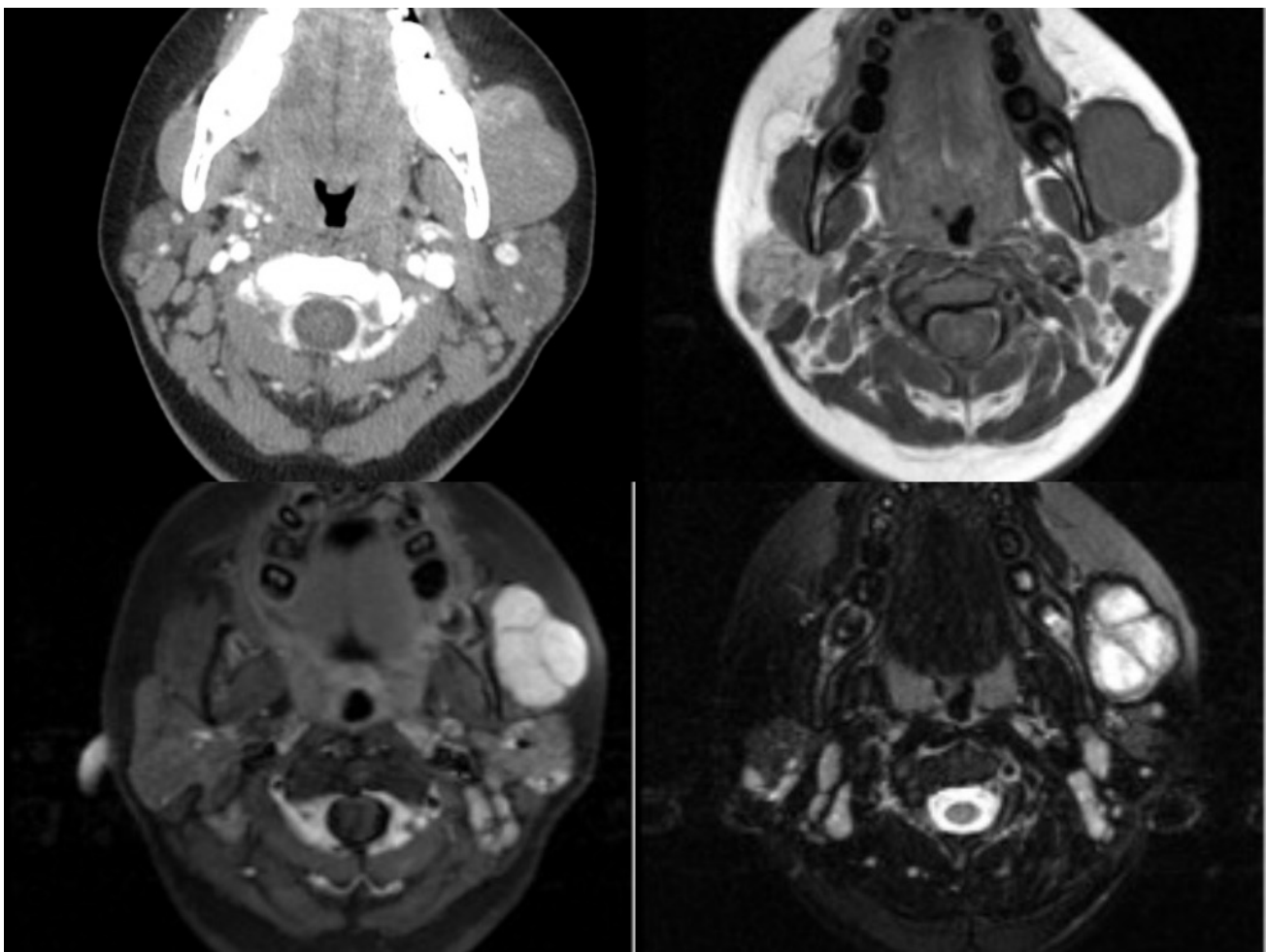


Figure 1: CT of the neck soft tissue with contrast, showing a mass in the left buccal area (Top Left). Axial T1-weighted MRI of the head and neck, showing a mass on left with isointense signal (Top Right). T1-weighted MRI with fat suppression, showing the nodular organization of the mass (Bottom Left). T2 weighted MRI with fat suppression, showing the nodular organization of the mass in addition to a narrow rim around the nodules resembling the target shape description reported in the literature (Bottom Right).

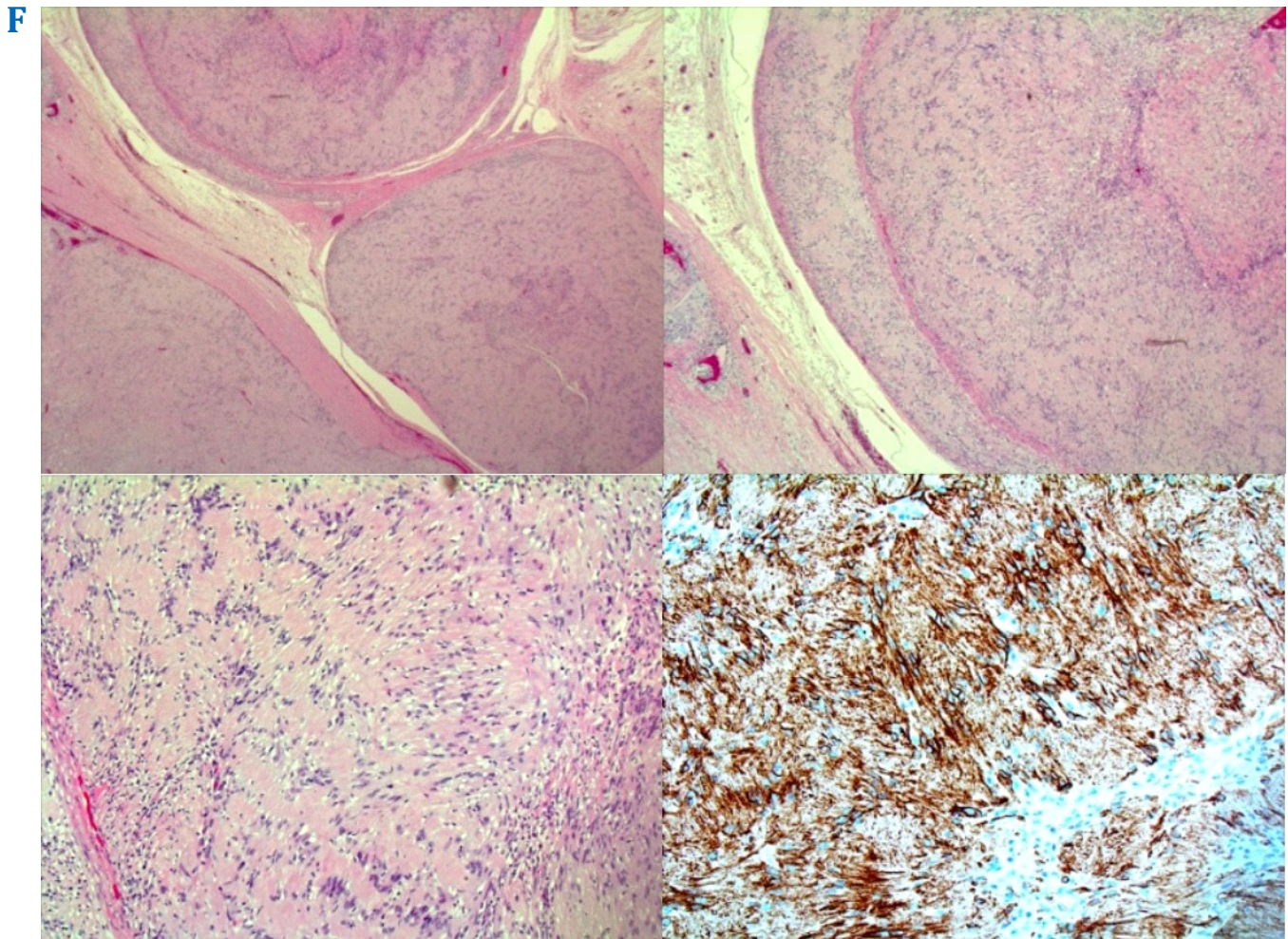


Figure 2: A 20X magnified micrograph of the tumor stained with H & E. this picture clearly demonstrates the multinodular organization of the schwannoma, hence the diagnosis of plexiform schwannoma (Top Left). H & E stained micrograph of the tumor with 40X magnification. Hyper cellular areas of organized spindle-shaped cells, also known as Antoni A areas, are evident in this picture (Top Right). 100X magnification showing a Verocay body and nuclear palisading (Bottom Left). An immunohistochemical staining of the tumor with CD56 in 200X magnification that shows prominent staining with the CD56 marker (Bottom Right).

References

1. Putney FJ, Moran JJ, Thomas GK. Neurogenic tumors of the head and neck. *Laryngoscope*. 1964; 74:1037-1059
2. Das Gupta T, Brassfield R, Strong E. Benign solitary schwannoma (neurilemmoma). *Cancer* 1969; 24:355-66
3. Harkin JC, Arrington J, Reed R. Benign plexiform schwannoma; a lesion distinct from plexiform neurofibroma. *J NeuropatholExpNeurol* 1978;37:622
4. Rongioletti F, Drago F, Rebora A. Multiple cutaneous plexiform Schwannomas with tumors of the central nervous system. *Arch Dermatol* 1989; 125 (3) : 431-432
5. Berg JC, Scheithauer BW, Spinner RJ, et al. Plexiform schwannoma: a clinicopathologic overview with emphasis on the head and neck region. *Hum Pathol* 2008; 39: 633-640
6. Wippold II FJ, Lubner M, Perrin RJ, et al. Neuropathology for neuroradiologist: A ntoni A and Antoni B tissue patterns. *Am J Neuroradiol* 2007; 28: 1633-1638
7. Fine SW, McClain SA, Li M. Immunohistochemical staining for calretinin is useful for differentiating schwannomas from neurofibromas. *Am J ClinPathol*. 2004 Oct;122(4):552-9

8. Hirose T, Tani T, Shimada T, et al. Immunohistochemical demonstration of EMA/Glut1-positive perineurial cells and CD34-positive fibroblastic cells in peripheral nerve sheath tumors. *Mod Pathol.* 2003 Apr;16(4):293-8
9. Lee DW, Byeon HK, Chung HO, et al. Diagnosis and surgical outcome if intraparotid facial nerve schwannoma showing normal facial nerve function. *Int J Oral Maxillofac Surg* 2013; 42:874-879
10. Mehta RP, Deschler DG. Intraoperative diagnosis of facial nerve schwannoma at parotidectomy. *Am J Otolaryng* 2008; 29: 126-129
11. Hebert-Blouin MN, Amrami KK, Scheithauer BW, et al. Multinodular/plexiform (multifascicular) schwannomas of major peripheral nerves: an underrecognized part of the spectrum of schwannomas. *J Neurosurg* 2010; 112: 372-382
12. Shimizu, K Iwai H, Ikeda K, et al. Intraparotid facial nerve schwannoma: A report of five cases and an analysis of MR imaging results. *Am J Neuroradiol* 2005; 26:1328–1330
13. Woodruff JM, Marshal ML, Goodwin Ta, et al. Plexiform (multinodular) schwannoma: A tumor simulating the plexiform neurofibromatosis. *Am J SurgPathol* 1983; 7 : 691-697
14. Rodgers GK, Myers EN. Surgical management of the mass in the buccal space. *Laryngoscope* 1988; 98:749-753
15. Battoo AJ, Sheikh ZA, Thankappan K et al. Nerve-sparing subcapsular resection of head and neck schwannomas: technique evaluation and literature review. *J LaryngolOtol* 2013 Jun; 4:1-6

Manuscript Information: Received: June 29, 2015; Accepted: August 25, 2015; Published: August 29, 2015

Authors Information: Soroush Farnoosh, BS^{a*}; Debra Don, MD^b; Masoud Shamszadeh, MD^c; Jeffrey Koempel, MD^b

^aUniversity at Buffalo School of Medicine and Biomedical Sciences. Buffalo, NY, USA.

^bDivision of Otolaryngology – Head and Neck Surgery, Children's Hospital Los Angeles. Outpatient Tower, First floor 4650 Sunset Blvd. Los Angeles, CA 90027, USA

^cDivision of Anatomical Pathology, Children's Hospital Los Angeles 4650 Sunset Blvd. Los Angeles, CA 90027, USA

Citation: Farnoosh S, Don D, Shamszadeh S, Koempel J. Plexiform schwannoma embedded within the masseter muscle: A case report. *Open J Clin Med Case Rep.* 2015; 1030

Copy right Statement: Content published in the journal follows Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>). © Farnoosh 2015

Journal: Open Journal of Clinical and Medical Case Reports is an international, open access, peer reviewed Journal focusing exclusively on case reports covering all areas of clinical & medical sciences.

Visit the journal website at www.jclinmedcasereports.com

For reprints & other information, contact editorial office at info@jclinmedcasereports.com