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Wagner-Meissner neurilemmoma of the right cheek

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Abstract We report a very rare case of Wagner-Meissner neurilemmoma in the cheek of a 10-year-old boy. The tumor presented as a slowly growing soft tissue swelling. Magnetic resonance imaging disclosed a very infiltrative, 9-cm mass involving the subcutis and deep soft tissues of the right cheek. Microscopically, the tumor was unencapsulated and composed almost entirely of well-formed Wagner-Meissner corpuscles that formed confluent sheets, perivascular cuffs, and individual corpuscles percolating through adipose tissue. Compared with the 3 previous reports, which describe circumscribed, encapsulated tumors in adult patients, this case had distinctive clinicopathologic features never reported: presentation in a pediatric patient, location in the head and neck region, and an infiltrative growth pattern.

Keywords: Wagner-Meissner; Meissner corpuscle; Neurilemmoma

1. Introduction

Wagner-Meissner corpuscles are specialized end organs that cushion the terminations of rapidly adapting afferents of mechanoreceptive neurons in glabrous skin. The rapidly adapting afferents are the neurons, which have long been known to be responsible for detecting low-frequency vibration. More recently, it has been proposed that they are also responsible for feedback signals important for grip control. Rapidly adapting afferents are particularly sensitive to minute, dynamic skin changes. Wagner-Meissner corpuscles are thought to protect these highly sensitive afferents from the confounding effects of large, static skin deformation [1,2].

Wagner-Meissner corpuscles are normally found in the dermis of the hands and feet, lips, tip of tongue, palpebral conjunctiva, and mammary nipples [3]. They are located in dermal papillae between sweat ducts and adhesive ridges in direct contact with the epidermal basal layer [2,4]. Approximately 2 to 9 rapidly adapting afferents from the subepidermal plexus terminate in a single Wagner-Meissner corpuscle [2]. These afferents demyelinate just before entering the corpuscle, and within the corpuscle, their axons branch repeatedly and form bulbous ribbon to disk-like enlargements. Axons wrap around the core of the corpuscle in a helical spring-like coil, and stacks of intervening lamellar modified Schwann cells are tightly bound to these spiraling axons. The bodies of Schwann cells containing nuclei localize peripherally in the corpuscle [5-7]. The distinctive lamellated Schwann cells of the Wagner-Meissner corpuscle give it a characteristic histologic appearance, which on routine light microscopy are round to oval encapsulated structures with central eosinophilic lamellations and peripherally oriented nuclei [8].

Wagner-Meissner corpuscles commonly comprise a minor component of several types of benign soft tissue tumors of neural origin; they occur as a secondary feature in neurofibromas and have been reported in traumatic neuromas and cellular nevi [9-12]. In 1986, Kaiserling and Geerts [12] first described a benign tumor, which was composed principally of Wagner-Meissner corpuscles. They coined this tumor *Wagner-Meissner neurilemmoma*. In this article, we present a novel case of a Wagner-Meissner neurilemmoma in the right cheek of a 10-year-old boy with distinctive pathologic features not previously described.

2. Clinical history and physical examination

The patient was a 10-year-old otherwise healthy African American boy who presented with an area of hyperpigmentation over his right cheek since birth. Over the 3 years

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before presentation, his parents noted that the hyperpigmented area had slowly enlarged and darkened with development of an underlying soft tissue mass. The soft tissue mass was occasionally tender after chewing but was otherwise asymptomatic. On physical examination, a generalized enlargement of the right cheek was noted, which extended to the infraocular cheek. Several overlying hyperpigmented macules and very flat papules were present. The mass appeared extremely soft, ill defined, and "doughy." Clinically, it was felt to be most consistent with lymphangioma, lymphatic malformation, or hemangioma. Neurofibroma was also in the clinical differential diagnosis, but was thought to be less likely. Imaging studies and biopsy of the mass were recommended.

3. Imaging

Magnetic resonance imaging (Fig. 1) of the skull base and upper neck showed an irregular, strongly enhancing soft tissue mass measuring 9 cm in anterior-posterior dimension, 5 cm in craniocaudal dimension, and 4 cm in maximum thickness. The mass was centered in the right masticator space superficial and deep to the anterior aspect of the masseter muscle extending along the fascia anteriorly to the nose. It extended both superficial and deep to the zygomatic arch and posteriorly toward the anterior aspect of the parotid gland. There was no evidence of tumor infiltration into the facial bones. Stranding of the subcutaneous tissue overlying the mass in the right cheek was identified. The inhomogeneous T1 and T2 signal and the partly inhomogeneous strong enhancement favored a lymphangioma or a lymphatic malformation over a hemangioma.

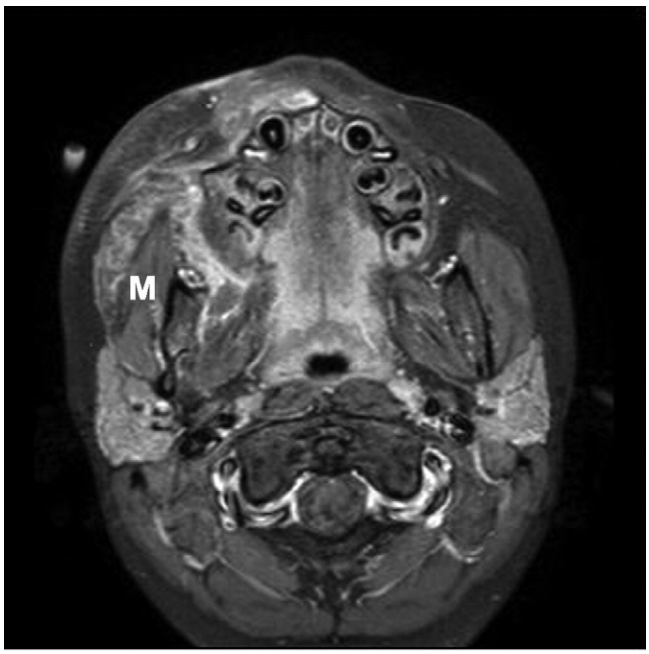

Fig. 1. Axial T1 postcontrast magnetic resonance image discloses an irregular, multilobulated enhancing mass in the right masticator space, both superficial and deep to the anterior aspect of the masseter muscle (M). Anterior extension of the mass is noted abutting the right maxillary alveolar process, whereas posterior extension abuts the parotid gland.

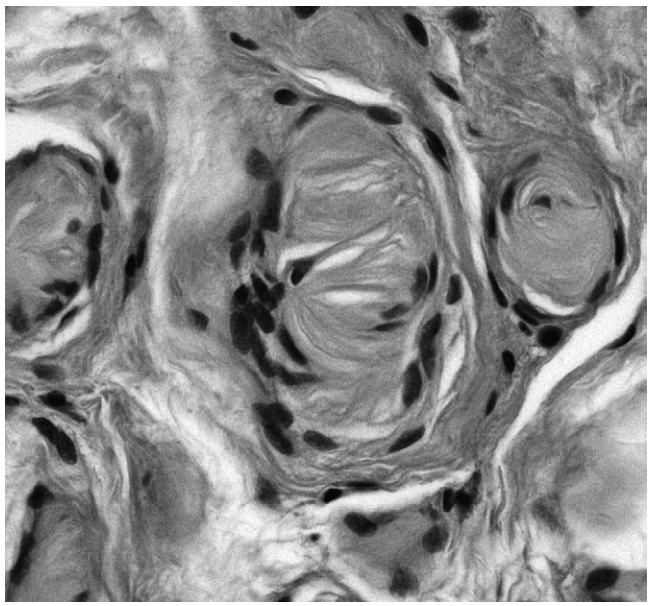

Fig. 2. The tumor was composed almost entirely of well-formed Wagner-Meissner corpuscles with characteristic central eosinophilic laminated cells and peripherally oriented nuclei.

4. Histology

The excisional biopsy of the mass was a 2.2 × 2.0 × 1.3-cm irregular soft tissue fragment with a cut surface that consisted of tan gray, glistening firm tissue admixed with adipose tissue. Histologically, a poorly circumscribed, unencapsulated tumor was present in the subcutis, with areas of tumor intimately admixed with adipose tissue. The tumor was composed entirely of Wagner-Meissner corpuscles that had characteristic 5 to 20 central eosinophilic lamellated cells with peripherally oriented nuclei (Fig. 2).

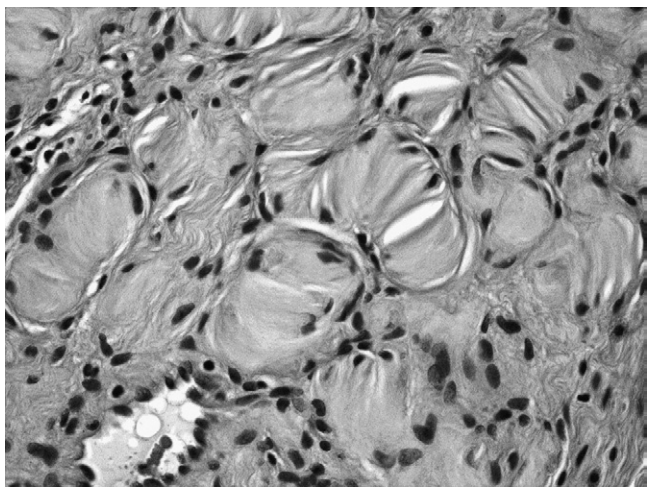

Fig. 3. In several areas of the tumor, Wagner-Meissner corpuscles became confluent, imparting a "paving stone-like" pattern.