Human Pathology (2017) 63, 70–78

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Human PATHOLOGY

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Original contribution

Clinicopathological features of peripheral nerve sheath tumors involving the eye and ocular adnexa☆,☆☆

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Received 12 December 2016; revised 16 January 2017; accepted 9 February 2017

Keywords:
Peripheral nerve sheath tumor;
Orbit;
Ocular;
Neurofibroma;
Schwannoma

Summary Peripheral nerve sheath tumors (PNSTs) are known to occur in the orbit and comprise 4% of all orbital tumors, but have not been well studied in contemporary literature. Ninety specimens involving the eye and ocular adnexa (1979-2015) from 67 patients were studied. The mean age was 32.5 years. Locations included orbit (58.9%), eyelid (60.0%), and other ocular adnexa. Most specimens were neurofibromas (70.0%), followed by schwannomas (11.1%), neuromas (11.1%), granular cell tumors (n = 4), nerve sheath myxomas (n = 2), and malignant PNST (n = 1). Fifty-six (88.9%) neurofibroma cases were neurofibromatosis 1 associated. Among neurofibromas, 31.7% were localized, 38.1% were plexiform, 25.4% were diffuse, and 4.8% were diffuse and plexiform. These tumors involved skin (31.7%), soft tissue (11.1%), skeletal muscle (22.2%), peripheral nerve (63.0%), lacrimal gland (20.6%), and choroid (n = 1). Other histologic findings included pseudo-Meissner corpuscles (27%), Schwann cell nodules (4.8%), prominent myxoid component (7.9%), melanin-like pigment (3.2%), and inflammation (14.3%). Available immunostains included S100 (+ in 15/15 cases), EMA (+ in 2/4 cases), CD34 (+ in 4/4 cases), and Ki-67 (<1% in 4/4 cases). Among 10 schwannomas, 8 were conventional and 2 were plexiform. Observed features included capsule (n = 5), hyalinized vessels (n = 5), Verocay bodies (n = 7), and Antoni B pattern (n = 5). Immunostaining included S100+ in 4 of 4 cases, and collagen IV+ and Ki-67 < 1% in 3 of 3 cases. Neurofibromas are the most common PNST involving the eye and ocular adnexa, and the majority are associated with neurofibromatosis 1. Plexiform and diffuse patterns and the presence of pseudo-Meissner corpuscles are relatively frequent in this area.
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☆ Competing interests: The authors have no financial disclosures or conflicts of interest.
☆☆ Funding/Support: This work was supported in part by a collaborative grant between the Wilmer Eye Institute (Baltimore, MD) and the King Khaled Eye Specialist Hospital (Riyadh, Saudi Arabia).
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http://dx.doi.org/10.1016/j.humpath.2017.02.006
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1. Introduction

Peripheral nerve sheath tumors (PNSTs) are common soft tissue and cutaneous neoplasms, but only comprise approximately 4% of all orbital tumors [1]. These tumors are thought to originate from sensory nerves in the orbit and are most frequently located in the superior and medial orbital compartments. The most common PNST of the orbit is neurofibroma [1,2], which can be classified as plexiform, diffuse, or localized; 2% of orbital tumors are plexiform neurofibromas frequently associated with neurofibromatosis type 1 (NF1), and 1% are isolated neurofibromas. The remaining PNSTs are schwannomas, the next most common type at this site. In addition, other rare PNSTs of the orbit such as neuroma, granular cell tumor (GCT), nerve sheath myxoma (NSM), and malignant PNST (MPNST) have been described.

To our knowledge, the largest case review of PNSTs of the orbit included 54 cases of neurofibromas, schwannomas, and MPNSTs; however, this study included only clinical information with no comprehensive pathologic analysis [3]. Neurofibroma-specific studies included a review of 13 patients with plexiform neurofibromas of the eye region with particular attention to cases in patients without NF1 [4] and multiple single-case reports [5-8]. Given their rarity, there have been small clinicopathological studies of orbital MPNSTs from 1985 (8 cases) [9] and 1989 (3 cases) [10], but only single-case reports in more recent literature [11-13]. Similarly, the largest report of 7 orbital schwannoma cases with histology was from 1982 [14], whereas only single-case reports have been published in the past decade [15-20]. Similarly, there has been a review of 19 cases of GCT in 1983 [21] and a literature survey performed in 2012 of 39 reported cases [22]. In contrast, we only found descriptions of a few cases of amputation neuromas [23,24] and one case report of an orbital NSM with histopathologic examination [25].

Thus, in recent literature, orbital PNSTs have not been well studied and have primarily been described on a clinical case-by-case basis. In this study, we performed a comprehensive histopathologic review of all orbital PNST cases seen at our institution between 1979 and 2015, and classified them using contemporary diagnostic histopathologic criteria.

2. Materials and methods

Pathology reports from the Eye Pathology Laboratory and the electronic Pathology Data System of The Johns Hopkins Hospital were searched for PNSTs involving the orbit and ocular adnexa, with identified specimens spanning from 1979 to 2015. The following search terms were used: neurofibroma, schwannoma, nerve sheath myxoma, plexiform, neurofibrosarcoma, MPNST, neuroma, neurothekeoma, perineurioma, and granular cell tumor. Surgical pathology reports were individually reviewed to identify only cases involving the orbit and ocular adnexa. Clinicopathological information (including patient age, sex, race, and medical history) and follow-up information regarding clinical, imaging, operative, and surgical pathology reports were recorded, if available. All histopathologic slides and special stains performed were reviewed, and their morphologic features were recorded. The study was approved by the institutional review board of Johns Hopkins Medicine, and all the recommended ethical guidelines were followed.

2.1. Statistical analysis

Continuous variables were compared using 1-way analysis of variance. *P* values less than .05 were considered statistically significant. Statistical analyses were performed using Stata 12.1 (StataCorp, College Station, TX).

3. Results

3.1. Patient demographics and history

A total of 90 specimens from 67 patients were identified (Table 1). The mean (SD) age was 32.5 (24.8) years, and 52.2% were female (47/90). Fifty-one (56.7%) of 90 were white, and 26 (28.9%) were black. The most common location of PNSTs was the orbit (37.8%), followed by the eyelid (36.7%) and both the eyelid and orbit (18.9%). Forty-nine (54.4%) of 90 tumors were surgically removed via gross total resection (GTR), 31 (34.4%) were removed via subtotal resection (STR), and 7 (7.8%) were biopsies. The mean (SD) recurrence-free survival (RFS) of GTR tumors, defined as the number of years between the operation date and the first tumor recurrence or death, was 5.3 (5.0) years. The minimum RFS was 0.25 years, and the maximum RFS was 16 years, which was a neuroma case where the patient died of reasons unrelated to the tumor.

Of all the PNSTs, 70.0% (63/90) were neurofibromas, 11.1% (10/90) were schwannomas, 11.1% (10/90) were neuromas, 4.4% (4/90) were GCTs, 2.2% (2/90) were NSMs, and 1 case was an MPNST. A total of 63 specimens from 42 patients were neurofibromas. The mean (SD) age of the neurofibroma specimens was 26.5 (20.9) years; 49.2% (31/63) were female, 57.1% (36/63) were white, and 31.7% (20/63) were black. An equal number of neurofibromas were located in the orbit and eyelid (33.3% each), with 27.0% of tumors extending through the eyelid and the orbit. Similarly, an equal number were excised via GTR and STR (44.4% each), and 6.3% were biopsies. The mean (SD) RFS of patients with a GTR neurofibroma was 4.5 (4.2) years. The minimum RFS was 0.25 years, and the maximum was 14.5 years; 22.2% (14/63) of patients are alive without tumor recurrence. In the neurofibroma subgroup, there was no significant difference in RFS with respect to the neurofibroma subtype ($P = .13$).

3.2. Neurofibromas

The morphologic features of neurofibromas are summarized in Table 2. Examples of these features are shown in

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