Rhabdomyosarcoma of the Orbit
A Clinicopathologic Study of 55 Cases

By
J. F. PORTERFIELD* and L. E. ZIMMERMAN**

With 8 Figures in the Text

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Rhabdomyosarcoma of the orbit, once considered a rare tumor, has been recognized with increasing frequency during the past few years. As a matter of fact, recent experience at the Armed Forces Institute of Pathology (ZIMMERMAN) and elsewhere (FRAYER and ENTERLINE) suggests that this is the most common malignant orbital tumor occurring in childhood.

In a review of the literature to 1942, CALHOUN and REESE could find only 14 reported cases of orbital rhabdomyosarcoma, to which they added 5 of their own. STOUT (2), who by September 1948 had studied 35 cases, reported only 1 from the orbit. In 1950 STOBBE and DARGEON reported 15 cases of embryonal rhabdomyosarcoma of the head and neck; 3 of the tumors were primary in the orbit. Nine years later, MOORE and GROSSI added 37 more rhabdomyosarcomas of the head and neck regions, 8 of which arose in the eyelid or orbit.

HORN and ENTERLINE, in 1958, reported 39 cases of rhabdomyosarcomas from all anatomic locations. Five of their tumors were primary in the orbit. BLAXTER and SMITH found only 33 cases of orbital rhabdomyosarcoma reported in the literature to 1958, to which they added 2 of their own. FRAYER and ENTERLINE reported 12 embryonal rhabdomyosarcomas of the orbit in 1959. The present series of 55 cases of orbital rhabdomyosarcoma not only is the largest yet reported, but it doubles the number of published cases.

Materials and Methods

The present report is an outgrowth of a clinicopathologic study of over 1,000 orbital tumors on file in the Registry of Ophthalmic Pathology at the Armed Forces Institute of Pathology (PORTERFIELD). Among these we have found 55 examples of orbital rhabdomyosarcoma with sufficient data to include in this study.

Based on their histopathologic characteristics, each rhabdomyosarcoma was classified into one of three histologic types: embryonal, differentiated, or alveolar. The histologic features of the embryonal and alveolar types have been described in detail by other investigators [ENTERLINE and HORN; ENZINGER; HORN and ENTERLINE; RIOPELLE and THERIAULT; SHUMAN; STOBBE and DARGEON; STOUT (2)], whose criteria for classification have been utilized in the present study. We have added a “differentiated type” to our classification, anticipating that the more highly differentiated neoplasms might have a better prognosis.
In some instances there was a mixture of histologic types; such cases were classified according to the predominant pattern. A "pleomorphic type" of rhabdomyosarcoma, which is usually located in the peripheral musculature of adults, has been described (Horn and Enterline). A purely pleomorphic tumor comparable to theirs was not recognized in our cases, but there was a certain pleomorphism in many of our tumors.

In each case, sections were stained with hematoxylin-eosin. When adequate material was available, the following additional stains were prepared: Masson's trichrome, periodic acid-Schiff reaction (with and without diastase), Mallory's phosphotungstic acid hematoxylin, and Wilder's reticulum (Manual of histologic and special staining techniques).

Clinical and follow-up data were correlated with each of the three histologic types. These data will be discussed separately with each histologic type.

Results

A. Embryonal Type (40 Cases)

Clinical Features. All patients with the embryonal type of rhabdomyosarcoma were Caucasian. The youngest was a newborn baby; the oldest 19 years. Their average age was 8 years (Table 1). Twenty-five were male, and 15 were female. The most common presenting sign was rapidly developing proptosis (Fig. 1). Proptosis was present in 31 and absent in 3; no information concerning it was available in 6 of the 40 cases. With one exception, the displacement of the eye was in a downward and temporal direction in the 19 cases in which the direction of proptosis was described. The direction of proptosis corresponds well with the most frequently stated location of these tumors—the upper inner quadrant of the orbit.

Less constant clinical signs and symptoms were palpable subconjunctival mass, redness of the eye, edema of the lids and conjunctiva, and ptosis of the lids. Pain was rarely an early symptom. Except for an orbital soft tissue density, results of roentgenographic studies were negative.

The tumors were described variously as soft, myxoid, grayish-white to yellowish, poorly defined masses in the upper inner quadrant of the orbit. In many instances the lesion extended into one of the eyelids, usually the upper lid. In no instance did the surgeon describe a tumor arising from one of the extraocular muscles. Except in a few cases in which no description was provided, the tumor was said to arise in the connective tissues, not in the extraocular muscles. At least two cases presented a multinodular subconjunctival mass similar to the sarcoma botryoides of the genitourinary tract. Since the term "botryoid" describes a gross characteristic, however, and since our two examples did not differ histologically from others classified as "embryonal", these cases were placed in the latter category.

Histologic Features. Although the histologic picture of embryonal rhabdomyosarcoma is fairly characteristic, there are many variations. Typically, stellate and spindle cells are arranged in a loose syncytium (Fig. 2a). In other areas, the syncytial pattern is less obvious, and the cells are more closely packed. Fre-