

Retrobulbar granular cell myoblastoma

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Granular cell myoblastoma is a relatively uncommon lesion of debatable histogenesis, appearing as a small, solitary (but occasionally multiple) neoplasm and usually taking a benign course. The malignant counterpart has rarely been observed (Ross, Miller, and Foote, 1952; Gamboa, 1955). Most granular cell myoblastomas have been observed in tongue, skin, and subcutaneous tissue (Crane and Tremblay, 1945; Strong, McDivitt, and Brasfield, 1970) and in the breast (Friedman and Hurwitt, 1966; Greenberg, 1967); examples have also been found in virtually every organ or tissue of the body (Strong and others, 1970), but only rarely in the orbit.

Von Bahr (1938) reported an orbital granular cell myoblastoma situated in the region of the lacrimal sac. Dunnington (1948) observed one in the upper part of the orbit leading to proptosis and exophthalmos and one in the region of the lacrimal sac. Dhermy, Morax, and Jolivet (1966) reported one in the eyelid of a 30-year-old woman.

We have recently seen a retrobulbar granular cell myoblastoma leading to proptosis and exophthalmos in a 15-year-old boy.

Case report

A 15-year-old mulatto boy was admitted to the Cancer Hospital of Paraiba, Brazil, with exophthalmos of 2 months' duration.

Examination

There was proptosis and exophthalmos of the right eye with marked palpebral oedema. X-ray examination showed no orbital bone involvement, and x-ray examination of the chest was within the normal limits.

Operation

A retrobulbar mass was found to be firmly attached to the optic nerve without invading it. The tumour, which was well-defined and of a firm and elastic consistency, was completely removed. It measured 5.4 × 3.0 cm.

Microscopic examination

There was a proliferation of large, round polygonal cells (Fig. 1), with a small round vesicular to hyperchromatic nucleus and a pink cytoplasm with coarse granules (Fig. 2). The cells were arranged in clusters or cords and were occasionally separated by varying amounts of fibrous collagen tissue. Periodic acid-Schiff-positive cytoplasmic granules were seen, but there were no mitotic figures. In some fields the cytoplasmic granularity was condensed into amorphous acidophilic masses (Fig. 3), and nuclei with prominent acidophilic nucleoli were commonly seen.

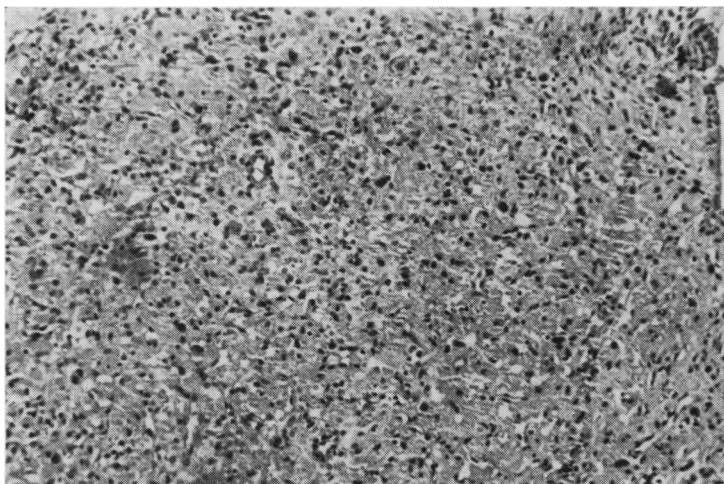


FIG. 1 *Proliferation of large polyhedral cells with coarse cytoplasmic granules. Haematoxylin and eosin. $\times 150$*

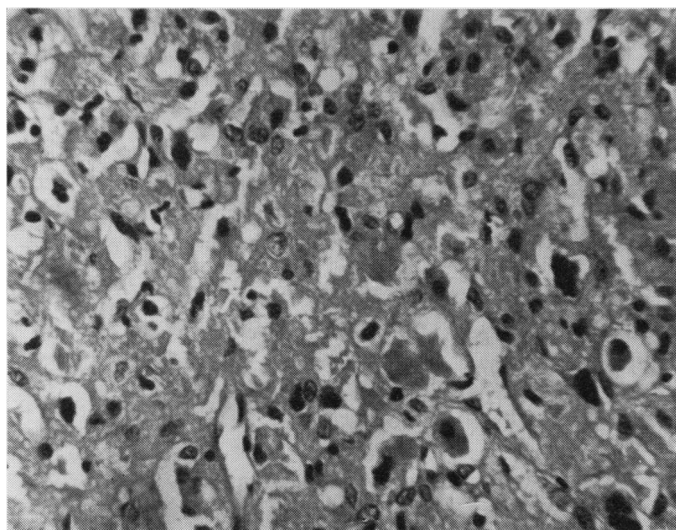


FIG. 2 *Groups of large cells containing many coarse granules in the cytoplasm and dense nuclear staining. Haematoxylin and eosin. $\times 300$*

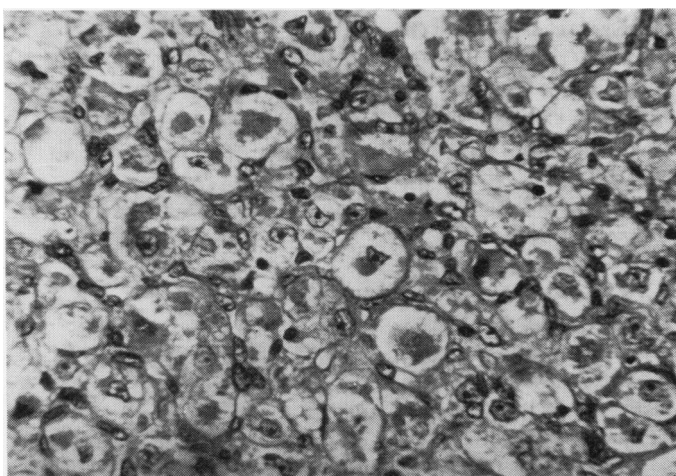


FIG. 3 *Concentrated hyalin masses within the cytoplasm. Note some cells with prominent nucleoli. Haematoxylin and eosin. $\times 400$*

Comment

Notwithstanding extensive histochemical, electron microscopic, biochemical, and clinical studies, the histogenesis of the granular cell myoblastoma remains controversial. The chief sites of origin are striated muscle (Abrikossoff, 1926; Murphy, Dockerty, and Broders, 1949; Murray, 1951), fibroblasts (Pearse, 1950), perivascular loose connective tissue (Toto and Restarski, 1967), and nervous tissue (Leroux and Delarue, 1939; Fust and Custer, 1949; Bangle, 1952; Ashburn and Rodger, 1952; Fischer and Wechsler, 1962). Some investigators relate the origin of the granular cell myoblastoma to a storage or metabolic derangement involving histiocytes (Gray and Gruenfeld, 1937; Azzopardi, 1956; Shear, 1960). According to Willis (1960), these lesions could be the result of injury to muscle fibres with subsequent degenerative or regenerative changes. The direct anatomical participation of the optic nerve sheath in the formation of the tumour in our case strongly suggests a neurogenic origin, probably related to the myelin sheath. Thorén (1950) found a biochemical similarity in the composition of the cytoplasmic granules of the granular cell myoblastoma to altered myelin as seen in experimentally damaged myelinated nerves. Histochemical studies also support the idea that the cytoplasmic granules originate from myelin (Pearse, 1950; Ashburn and Rodger, 1952; Fischer and Wechsler, 1962).

The only treatment is excision; the tumours are usually well encapsulated which makes their removal a simple procedure.

Summary

In a case of retrobulbar granular cell myoblastoma occurring in a 15-year-old mulatto boy, the tumour was firmly attached to the optic nerve sheath and caused proptosis. The authors suggest that it arose from the myelinated sheath of the optic nerve.

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