

Rhabdomyoma of the soft palate. Fine structural details of a highly differentiated muscle tumor

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Summary. An adult rhabdomyoma was light and electron microscopically studied. The lesional cells presented well-known structural details, such as abundance of mitochondria and glycogen, myofilaments and miofibrils, hypertrophied Z-bands and masses of Z-band material. Triads were randomly scattered in the cytoplasm and also related to the individual sarcomeres. In sarcomeres the triads were regularly placed near to the A-I-junctions. This peculiarity of mammalian skeletal muscle fibers may yield a criterion to distinguish between cardiac and extracardiac rhabdomyomas. Circumscribed surface areas of the tumor cells were provided with elaborate infoldings of plasma membrane and basal lamina. These areas were interpreted as imitating myotendinous junctions. Satellite cells were regularly found.

Key words: Adult rhabdomyoma — Extracardiac rhabdomyoma — Oral cavity — Electron microscopy

Introduction

In a recent review on tumors of skeletal muscle (Agamanolis et al., 1986) the authors indicate that about 2 per cent of skeletal muscle tumors represent rhabdomyomas. The rare lesions are further classified as adult, fetal, and genital rhabdomyomas (Enzinger and Weiss, 1983). Fine structural studies have shown the presence of thin and thick myofilaments, myofibrils with Z-discs, and triads in the tumor cells (Cornog and Gonatas, 1967). These morphological details of the tumor cells proper, together with the presence of satellite cells, indicate a high degree of differentiation, which is also reflected by the benign clinical course of the lesions. In the present paper we report on additional fine structural details

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observed in an adult rhabdomyoma of the soft palate. These observations concern surface differentiations bearing some resemblance to myotendinous junctions, and the spatial relationship between triads and myofibrils. The latter observation may be helpful to discern between cardiac and extracardiac rhabdomyoma.

Materials and methods

Case report.

In March 1981 a 72-year-old male was admitted to the municipal hospital at Lainz, Vienna, because of severe difficulty in breathing, which was caused by progressed lung emphysema. During inspection of the mouth a circumscribed tumorous lesion was observed in the soft palate. The soft, ovoid nodule measured about 25×40mm. It was located near to the right tonsil. After fine needle aspiration cytology the lesion was suspected to represent either an oncocytoma or tumor of myogenic origin. The nodule was surgically removed; it consisted of homogeneous brownish tissue that was well bounded by a connective tissue capsule and subdivided by delicate septa. The postoperative course was uneventful. 6 years after enucleation of the tumor there is no evidence of recurrence.

Smear preparations after fine needle aspiration were routinely stained **May-Grünwald/Giemsa** and **Papanicolaou**. For light microscopy tumor tissue was fixed in 5% formalin and embedded in paraffin. Sections were stained with **Hematoxylin/Eosin**, **PAS**, and Gomori's silver method for reticular fibrils. For electron microscopy small pieces of the tumor were fixed in 5% glutaraldehyde in phosphate buffer (pH=7.2) and postfixed in 1% osmium tetroxide in veronal acetate buffer (pH=7.2). Dehydration was done in graded ethanols and propylene oxide, embedded in Epon 812. Semithin sections were stained with alkaline Toluidine blue 0, thin section with methanolic uranyl acetate and lead citrate. Electron microscope: Zeiss EM9.

Results

A. Aspiration cytology

In the smear preparations large, polygonal to oval cells with strongly eosinophilic, faintly granular cytoplasm were conspicuous. Most of these cells contained a single nucleus in central position, a few of them were seen also to be accompanied by some peripherally located nuclei which could not clearly be assigned. Fibrillar inclusions or cross striations were not seen.

B. Light microscopy

The lesion was made up of round, oval, or moderately polymorphous cells, separated by thin fibrous septa. It was provided with capillaries in an otherwise scanty stroma. The large tumor cells showed abundant acidophilic granular cytoplasm, sometimes with fibrillar, cross striated inclusions. PAS staining revealed lakes of glycogen in the tumor cells and continuous basement membranes were seen around them. Argyrophilic reticular fibers were attached to the basal laminae. Only few of the neoplastic cells contained eosinophilic crystalline inclusions, presenting themselves as rods or tufts of needle-like crystals.

These cytological details of neoplastic muscle cells were seen much better in semithin sections from osmicated material stained with Toluidine blue (Fig. 1): cross striations were clearly demonstrated, lakes of glycogen presented as homogeneous, metachromatically-stained masses, the crystalline inclusions appeared as deeply basophilic rods or tufts, and the acidophilic granularity described in conventional sections was identified as

corresponding to masses of mitochondria (Fig. 1, a to d, respectively).

C. Electron microscopy

Most of the tumor cells contained an abundance of mitochondria and lakes of glycogen (Fig. 2). Mitochondria appeared as round or oval profiles with numerous transverse cristae; granulae mitochondiales were also apparent. In a few cases spherical mitochondria with concentrically oriented cristae were seen (Fig. 7). Glycogen deposits consisted exclusively of β -particles.

The neoplastic cells regularly contained abundant cytoplasmic filaments, both of the actin and myosin type.

These filaments showed a tendency to form rudimentary myofibrils composed of sarcomeres (Figs. 3, 4). Myofibrils were of varying length, often branched, an only rarely oriented in parallel to form the striated pattern of skeletal muscle fibers (Fig. 3). Z-band material was frequently seen isolated from sarcomeres, forming irregular masses or rod-like structures which most probably correspond to the crystalline inclusions seen in the light microscope. The sarcomeres were often accompanied by triads (Fig. 3, 4) but elements of the T- and L- systems were also found irregularly scattered throughout the cytoplasm. Triads were often seen in characteristic spatial relationship to sarcomeres at the boundaries between A- and I- bands (Fig. 4). Reduplication of triads were commonly noted (Fig. 3). All the neoplastic cells were provided with continuous basal laminae. The cell surface of tumor cells sometimes showed complex infoldings and invagination of plasma membrane and basal lamina (Fig. 5). Normally-looking satellite cells were regularly found (Fig. 6).

Fig. 1. Semithin section of the tumor shows tightly packed groups of polygonal cells separated by delicate strands of capillarized connective tissue (Cap). Characteristic inclusions are indicated: cross striated myofibrils (a), lakes of glycogen (b), basophilic rods and masses (c), and abundance of mitochondria (d). $\times 600$

Fig. 2. Low power electron micrograph shows neoplastic cells with numerous regularly built mitochondria (indicated d as in Fig. 1) and lakes of glycogen (indicated b as in Fig. 1). Septa of connective tissue with fibrocytes and capillaries (Cap.) are inconspicuous. Some of the tumor cells contain small lipid droplets. $\times 4,000$

Fig. 3. Region with regularly arranged myofibrils. Electron-dense material of Z-discs in abundant and shows a tendency to form clumsy masses which are not well-positioned at all (indicated c as are indicated masses of Z-disk material in Fig. 1). Triads are numerous. Sometimes these structures are composed to form «pentads»

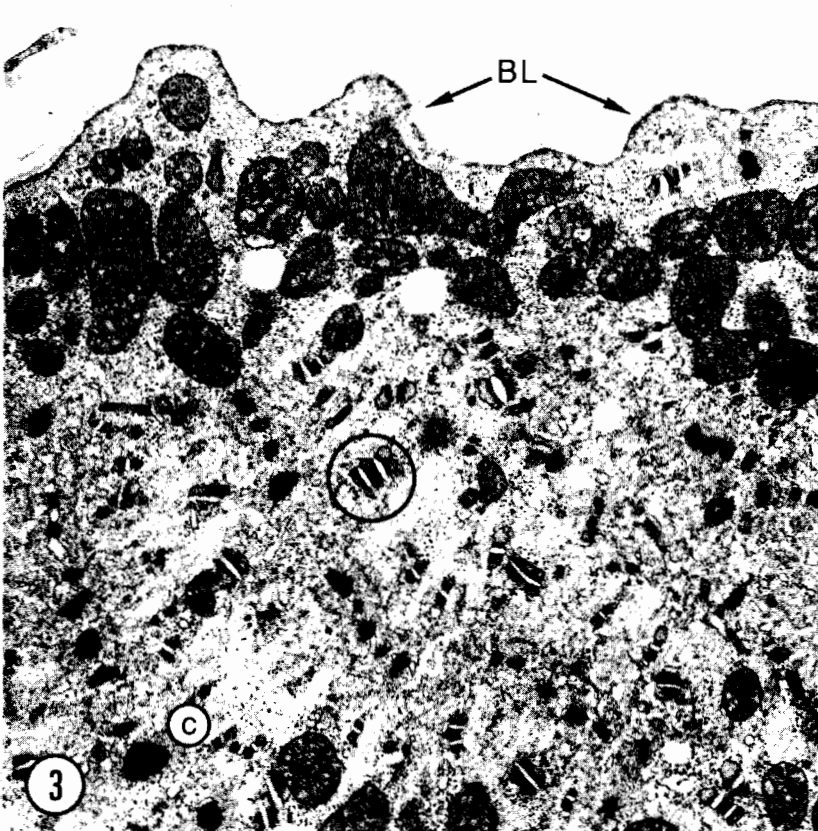
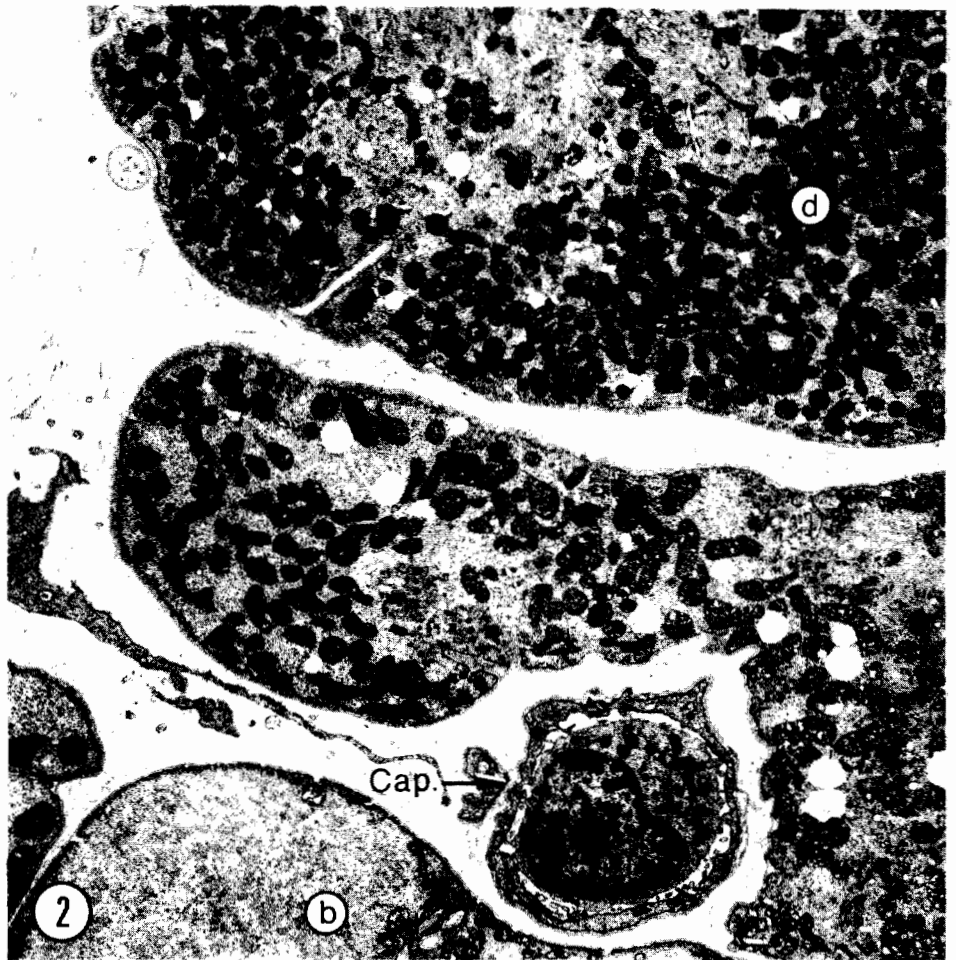
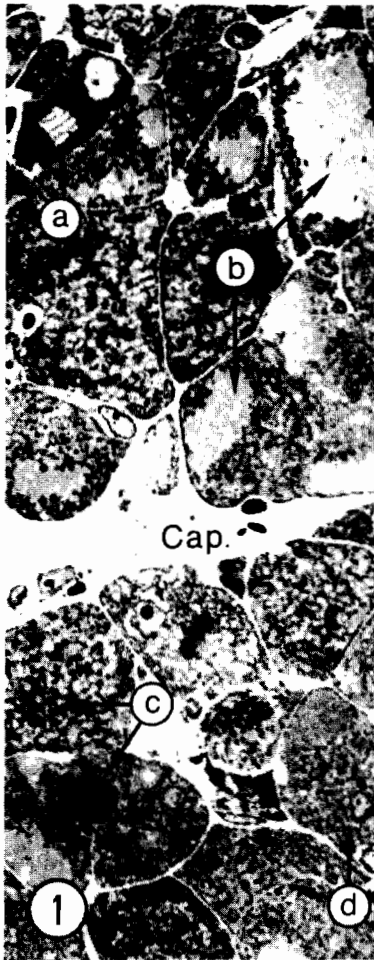
(encircled region). BL: basal lamina. $\times 12,000$

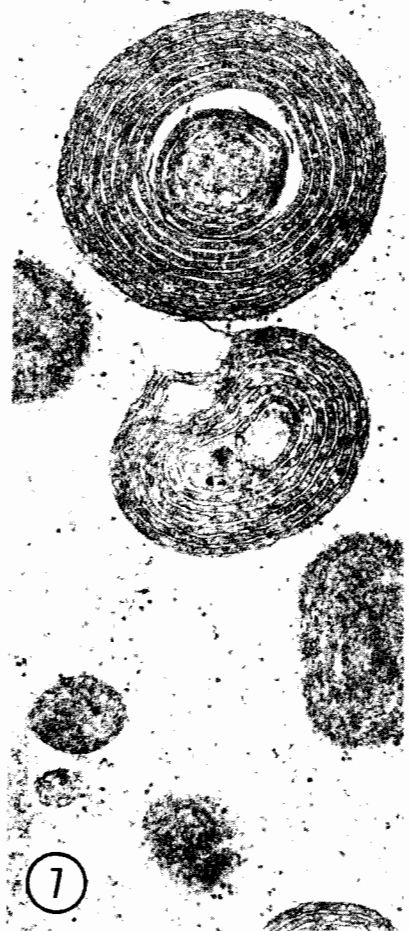
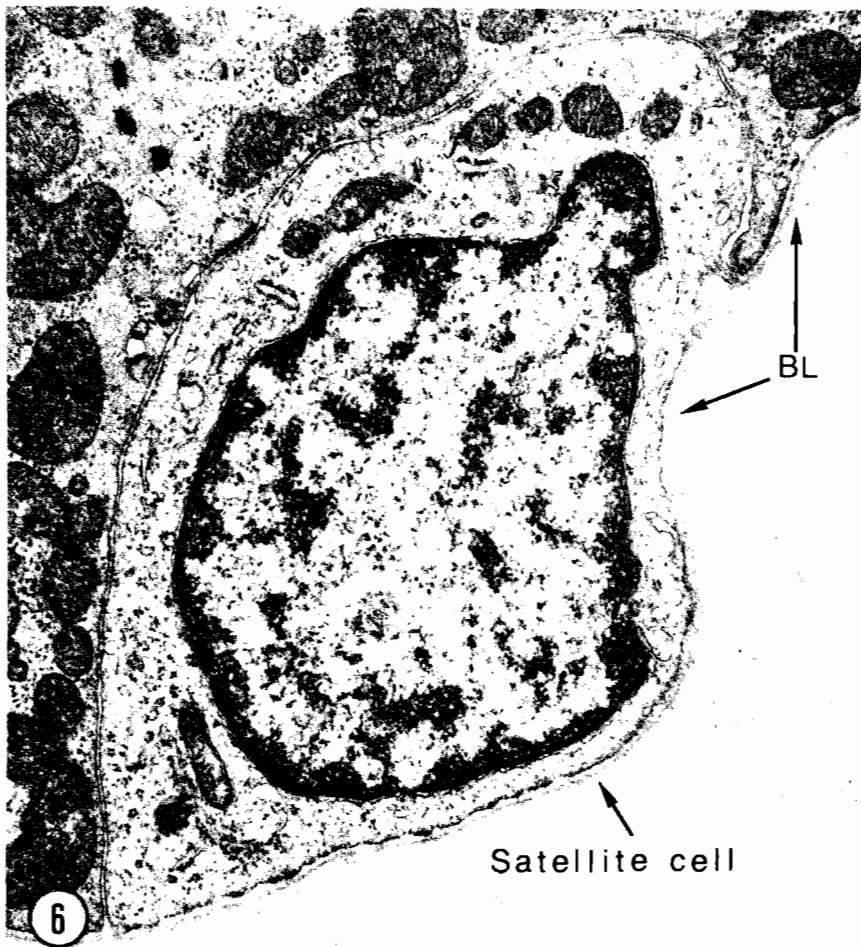
Fig. 4. Triads are regularly positioned at the A-I-junctions in the sarcomeres. Glycogen stores consist exclusively of β -particles. $\times 28,000$

Fig. 5. Fording and invagination of plasma membrane and basal lamina in circumscribed areas of the tumor cells bear some resemblance of the cellular part of myotendinous junctions. BL: basal lamina. $\times 28,000$

Fig. 6. Regularly built satellite cells are commonly seen to accompany the rhabdomyoma cells. BL: basal lamina. $\times 28,000$

Fig. 7. Figure shows mitochondria with concentrically arranged crista, which were observed preferentially in tumor cells crowded with glycogen. $\times 28,000$





Discussion

Light and electron-microscopical observations in the reported case of adult rhabdomyoma fully correspond to the morphological characteristics listed in recent reviews (Agamolis et al., 1986) or in basic original papers (Cornog and Gonatas, 1967; Tandler et al., 1970). Also, the localization of the tumor, age and sex of the patient, and clinical course of this case is characteristic (Heiden et al., 1978). Electron microscopy revealed an abundance of relatively small mitochondria in the neoplastic cells, which accounts for their misinterpretation as oncocytes in smear preparations. Mitochondria also cover over fibrillar or crystalline inclusions in the smears; these inclusions could readily be seen in sections. Even when reexamining the smears after having diagnosed the nature of the lesion, it was impossible to discern any inclusions.

Two fine structural peculiarities deserve further comment. First, some of the tumor cells showed an elaborate system of folds and invaginations of the plasma membrane and basal lamina (Fig. 5) A comparable situation has been shown by Gold and Bossen (1976; their Fig. 9) in a vaginal rhabdomyoma, but the authors did not further take notice of it. In our view these labyrinths resemble areas similar to those seen in myotendinous junctions (Hanak and Böck, 1971) and therefore may be regarded as an indication of differentiation. Second, triads are often positioned near to the boundary regions between A- and I- bands of the sarcomeres (Fig. 4), the triads are clearly not related to the Z-disks. This position of triads is characteristic for mammalian skeletal muscle, whilst the triads are related to Z-disks in heart muscle cells. We therefore interpret the spatial relationship of triads to A-I-junctions in the rhabdomyoma cells as an indication of its skeletal muscle origin. Cardiac

rhabdomyomas, on the other hand, should present diads or triads next to the Z-disks if there are any T-tubules and related structures at all. Unfortunately, cardiac rhabdomyoblasts are less well-differentiated as compared to extracardiac rhabdomyoma cells and T-tubules are often missing (Fenoglio et al., 1976; Silverman et al., 1978), as is also the case in normal Purkinje cells, specialized conducting cells and many atrial muscle cells.

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