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Case Report

Heterotopic Striated Muscle on the Surface of the Brain

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Summary. Heterotopic tissue, identified by both light and electron microscopy as cardiac muscle, was detected at autopsy on the surface of the right superior temporal gyrus of a 73-year old female patient. The cells of the heterotopic tissue were interconnected by intercalated discs and the fibres contained regularly cross-striated, contracted, or relaxed myofibrils indicating a continuing contractile activity until the time of death.

Key words: Heterotopic tissue – CNS – Cardiac muscle.

Introduction

Muscle tissue was detected on the surface of the right superior temporal gyrus by light microscopic examination of brain autopsy material from a 73-year-old female patient. Occurrence of muscle tissue in the central nervous system has been previously reported only in neoplastic diseases. Cross-striated and or smooth muscle elements were detected by Eberth (1898) in a subdural teratoma, by others (Russel and Rubinstein, 1971; Ingraham and Bailey, 1946; Leedham, 1972) in a teratoid variety of medullo-blastoma, the so-called medullo-myoblastoma. Shuangshoti et al. (1968), and Shuangshoti and Phonprasert (1976) reported two cases of rhabdomyosarcoma occuring in the cerebellum, and subfrontal region respectively. In the present case the muscle tissue was clearly non-neoplastic; electron microscopic examination revealed the presence of regular myofibrils and intercalated discs within the muscle fibres. Since occurrence of heterotopic non-neoplastic muscle tissue in the brain appears to be unprecedented in the literature, a description of the light and electron microscopic findings seems to be warranted.

Case Report

Mrs. H.F., 73 years old, was admitted to the 3rd Clinic of Internal Diseases, Postgraduate Medical School, Budapest, following the diagnosis of a cerebrovascular event which resulted in left hemiple-

gia. During hospitalization she developed fever, which was attributed to an urinary tract infection. Subsequently, respiratory and circulation insufficiency occurred and she died in 2 weeks.

At post mortem examination advanced cerebral sclerosis and concomitant softening were found. Changes indicative of chronic circulatory insufficiency were also present in the viscera, and acute pulmonary and cerebral oedema were found. The urinary tract infection had apparently been cured by treatment. Pathohistologically chronic interstitial myocarditis of the infectious allergic type was detected, which was the basic disorder responsible for death. Specimens from all inner organs were examined by light microscopy.

The *heterotopic tissue* was detected during the routine pathohistological examination of the brain. It was located in the right superior temporal gyrus, on the surface facing the ascending branch of the lateral sulcus, and was 0.5 mm in diameter. Fibres of 10 μm in diameter, reminiscent of *cardiac muscle,* were seen in the light microscopic specimens. The muscle tissue was surrounded by a connective tissue capsule, intimately interconnected with the surface of the brain. The encapsulated muscle fibres were practically embedded in the superficial layer of the cerebral cortex (Fig. 1). The outer surface of the heterotopic tissue, facing the meninges, was disrupted on peeling off the arachnoid layer. In parts of the muscle fibres, cross-striation was detected by several different staining techniques (H.E., van Gieson, PTAH). The PAS reaction revealed the presence of lipofuscin-like granules in many places, particularly in the perinuclear sarcoplasm. Some fibrous stroma was found between the fibres on staining with van Gieson's technique.

Following detection of the heterotopic tissue by light microscopy, a search was made for further similar islets by the naked eye, by stereomicroscopy and by light and electron microscopy, but none were found.

Subsequently part of the stained paraffin-embedded sections were post-fixed in 1% OsO_4, dehydrated in step-graded ethanol, and embedded in Araldite. Ultra thin sections were cut at right-angles to the plane of the original paraffin sections, stained with uranyl acetate, and counter-stained with lead citrate.

Fig. 1. Low power light micrograph of heterotopic muscle tissue (*HM*) associated with the cerebral cortex (*CC*). The two kinds of tissue are interconnected by a fibrous capsule (*C*). Paraffin section, H.E. × 140

Fig. 2. This specimen was originally treated using the PAS reaction, and the electron micrograph was taken in the plane of its cross section. Note cross-striation on the myofibrils. × 12,500

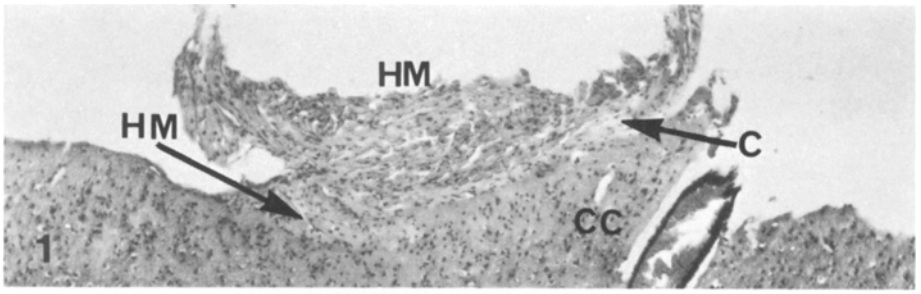
Fig. 3. Intercalated disc (*ID*) in one fibre of the heterotopic tissue. Note mitochondria near myofibrils at right. × 10,000

Fig. 4. Desmosome found in an intercalated disc. × 18,000

Fig. 5. Straight, markedly electron dense cristae (*arrows*) in the mitochondria of the heterotopic tissue. × 10,000

Fig. 6. Relaxed sarcomeres. The bands of the cross-striation are labeled. × 37,000

Fig. 7. Myofibrils in the state of maximal contraction. Only the Z, A and M bands are seen in the sarcomeres × 27,000


HM
HM
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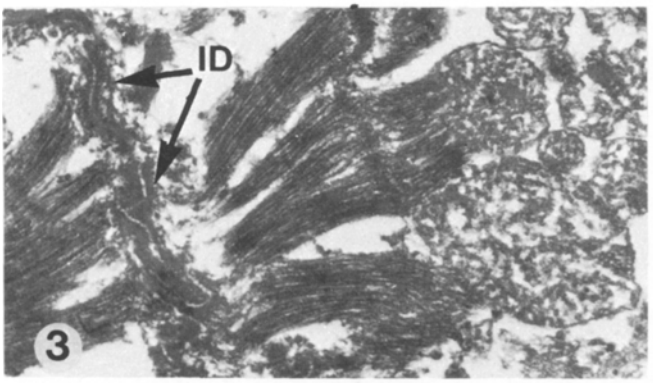
1


2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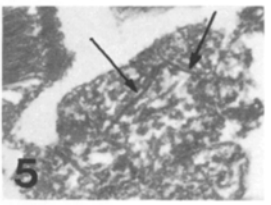
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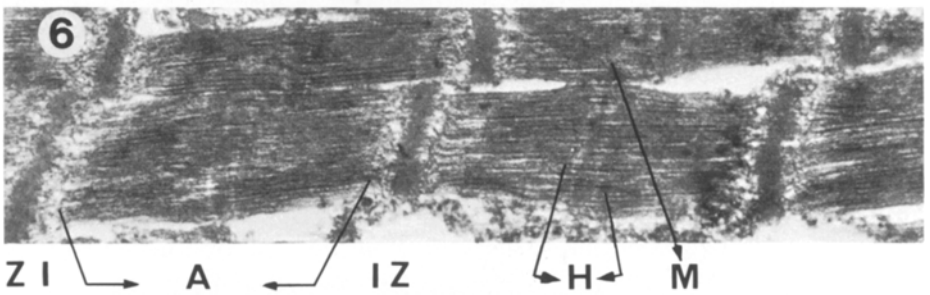
6
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A
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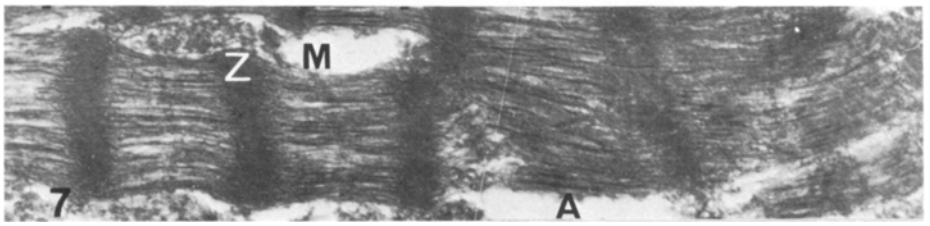
H
M


Z
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7
A

Somewhat surprisingly, neither the post-mortem changes, nor the double processing, affected the suitability of the specimens for electron microscopic study (Fig. 2). The cells of the fibres were interconnected by undulating intercalated discs, inside which the fascia adherens and desmosomes with myofibrillar and tonofibrillar attachments could be easily distinguished (Figs. 3 and 4). Although the mitochondria showed postmortem alteration, in some of them inner membranes reminiscent of those seen in ultracentrifuged myocardial preparations (Hall and Crane, 1970) and of those found in hepatocellular mitochondria (presumably under hypoxic conditions, Bartók et al., 1973) were seen (Fig. 5). Part of the myofibrils were in a state of maximal contraction, while others were relaxed, showing regular bands of cross-striation. Thin and thick filaments, I and A bands, as well as M and H bands, were easily distinguishable in the sarcomeres (Figs. 6 and 7). The tubules of the sarcoplasmic reticulum were in a state of almost complete lysis, but in places structures corresponding to triads remained at the margin of I and A bands. The central elements of the triads corresponded to tubular profiles 600–700 Å in diameter. Lipofuscin-like granules were dispersed between the myofibrils.

Discussion

Both light, and more particularly, electron microscopic examination demonstrate that the heterotopic tissue found on the surface of the right superior temporal gyrus was muscle tissue. The architecture of the tissue, and especially the presence of intercalated discs suggest that the heterotopic tissue is *cardiac* muscle, although triad remnants of sarcoplasmic reticulum at the borders of I and A bands are reminiscent of the SR organisation found in skeletal muscle (Sommer and Johnson, 1969). Since the heterotopic tissue was interconnected with the brain by a connective tissue capsule, the possibility of displacement of cardiac tissue during autopsy can be eliminated. In contrast to previous reports on the occurrence of muscle elements in neoplastic tissue (see Introduction), this heterotopic tissue was definitely non-neoplastic.

One possible explanation of this occurrence is that the cross-striated muscle tissue became associated with the brain during embryonic life, in the form of an aberrant mesenchymal element. Shuangshoti and Phonprasert (1976) suggested also that the intracranial rhabdomyosarcoma of their case arose from aberrant, unstable mesenchymal tissue which differentiated toward striated neoplastic muscle.

Differentiation of the neuroectoderm to cross-striated muscle tissue has been shown to occur in the avian embryo. The striated iridic sphincter muscle of the chicken derives from the ectoderm, and the fibres show a cross-striation as regular as that found in the heterotopic tissue in the present case (Brini et al., 1964; Porte, 1966).

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