Ecchordosis physaliphora

An electron microscopic study

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Ecchordosis physaliphora, an asymptomatic gelatinous excrescence arising from the clivus, is occasionally encountered at autopsy. Morphologically, ecchordosis closely resembles notochord and chordoma. All three consist of vacuolated cells that contain acid mucopolysaccharides. Under the electron microscope, the intracellular vacuoles have a smooth limiting membrane whereas the extracellular vacuoles are lined by microvilli and pinocytotic vesicles. No ultrastructural features accounting for the biological difference between ecchordosis and chordoma are observed in this study.

KEY WORDS • ecchordosis physaliphora • chordoma • notochord • ultrastructure • hematoma • clivus

Ecchordosis physaliphora, a gelatinous hamartoma arising from the clivus, was first described by Virchow in 1857. Although Virchow considered it cartilaginous, Müller, in the following year, advanced the view that ecchordoses were derived from notochord, a concept that took almost 40 years to be generally accepted. Ecchordoses are almost always small and asymptomatic, and have been encountered six times in the last 1600 complete autopsies in this institution, an incidence consistent with that reported in other autopsy series. While the light microscopic characteristics have been described in detail and compared to those of chordoma and notochord, ecchordoses have not been studied by electron microscopy. In this report the ultrastructure of two ecchordoses is described.

Materials and Methods

Grossly, one of the ecchordoses appeared as a small gray-white gelatinous mass within the leptomeninges on the ventral surface of the pons adjacent to the basilar artery. This mass was attached by a short thin pedicle to a bony excrescence on the mid-clivus (Fig. 1 left). The gelatinous material covered the bony excrescence and extended through it into the marrow spaces of the basisphenoid. The second ecchordosis was recognized on the removed brain as a gelatinous mass in the leptomeninges on the ventral surface of the mid-pons.

For light microscopy, tissue from the two ecchordoses was fixed in either buffered formalin or picric acid-formalin and was embedded in paraffin. Sections were stained with hematoxylin and eosin, Masson’s trichrome, periodic acid-Schiff (PAS) with and without antecedent diastase digestion, toluidine blue, alcin blue, mucicarmine, phosphotungstic-acid-hematoxylin (PTAH), and Hale colloidal iron.

For electron microscopy, blocks of formalin and picric acid-formalin-fixed tissue were
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postfixed in osmium tetroxide and embedded in any epoxy resin. Sections 2 μ-thick were stained with paraphenylene-diamine for screening. From selected blocks, thin sections were cut with a diamond knife and stained with uranyl acetate and lead citrate before examination under an electron microscope.

Results

When examined by light microscopy, both of the ecchordoses consisted of sheets of cells having numerous intra- and extracellular vacuoles (Fig. 1 right). The contents of the vacuoles were granular and stained red with hematoxylin and eosin, pink with mucicarmine, blue green with alcian blue and Hale's colloidal iron, and metachromatically with toluidine blue. The magenta staining produced by periodic acid-Schiff reaction was unaffected by diastase digestion. Neither Masson's trichrome nor phosphotungstic acid hematoxylin stained the contents of the vacuoles.

Examination under the electron microscope showed that the two ecchordoses were similar and consisted of interdigitating cells that often had long tenuous processes (Fig. 2). Although some cells had only a few small vacuoles, most cells had either several large vacuoles or a single very large vacuole displacing the cytoplasm into a thin peripheral rim (Fig. 3). All intracytoplasmic vacuoles were limited by a single, smooth membrane (Figs. 2 and 3) and many contained sparse, finely granular osmophilic material. Although most of the vacuoles were close to the rough endoplasmic reticulum, only a few small vacuoles could be demonstrated in continuity with the cisterns. Other vacuoles were adjacent to cisterns of the smooth endoplasmic reticulum or the plasma membrane.

The sparsely vacuolated cells appeared to have more mitochondria and rough endoplasmic reticulum than the highly vacuolated cells. Single cisterns of rough endoplasmic reticulum often partially or completely encircled one or more mitochondria (Fig. 4). Although these encircled mitochondria were usually located adjacent to the nucleus, no continuity between the cisterns of rough endoplasmic reticulum and the external nuclear membrane could be demonstrated.

Fine filaments, approximately 75 A in diameter, were scattered randomly or aggregated into broad sheaves within the cytoplasm of both the sparsely and highly vacuolated cells. They were more conspicuous, however, in the sparsely vacuolated cells.

The extracellular space varied in extent and configuration. Often adjacent plasma membranes were separated by only a few hundred Angstroms, and occasionally desmosomes were present on the opposing membrane surfaces (Fig. 5 left). In other areas the extracellular space was greatly expanded forming irregular vacuoles (Figs. 2 and 5 left). Some of these spaces contained small aggregates of granular osmophilic material. Long tenuous cytoplasmic processes and irregular wedge-shaped microvilli projected into these extracellular vacuoles. Pinocytic or emeoeytic vesicles were scattered along the margins of these extracellular vacuoles and were occasionally found along the plasma membranes of closely applied cells. Occasionally, an especially long cytoplasmic process deeply invaginated an adjacent cell. When sectioned transversely, these processes appeared as intracellular islands of cytoplasm bounded by double membranes. Unlike the intracellular vacuoles, these structures were filled with myriads of filaments and had pinocytic vesicles along the limiting membranes (Fig. 5 right).

Discussion

Light microscopy reveals that ecchordosis physaliphora like chordoma and notochord consists primarily of sheets of vacuolated cells. The staining with alcian blue and the metachromasia with toluidine blue demonstrates that the vacuoles contain acid mucopolysaccharides. The ultrastructural characteristics of these tissues closely resemble one another. All three consist of a single type of cell that varies in degree of cytoplasmic vacuolization, ranging from sparsely vacuolated cells to "signet-ring" cells. The intracytoplasmic vacuoles of notochord and chordoma cells have been variously regarded as being derived from rough endoplasmic reticulum, smooth endoplasmic reticulum, or invaginations of extracellular space. In ecchordosis, all of the intracytoplasmic vacu-
Fig. 1. Left: Ecchordosis physaliphora as seen at autopsy arising from the mid-clivus (arrow). Right: Ecchordosis physaliphora as seen by light microscopy consists of vacuolated and nonvacuolated cells. Some of the vacuoles appear to be extracellular. Trichrome, × 600.

Fig. 2. Electron micrograph showing a sparsely vacuolated cell with microvilli and cytoplasmic processes extending into dilated extracellular spaces that comprise the extracellular vacuoles (ecv). Intracellular vacuoles (icv) are present in adjacent cells. Uranyl acetate and lead citrate, × 13,000.
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**FIG. 3.** Electron micrograph showing two cells with large intracytoplasmic vacuoles (icv) that contain fine, granular, osmophilic material. Uranyl acetate and lead citrate, ×7500.

**FIG. 4.** Electron micrograph showing a sparsely vacuolated cell containing mitochondria encircled by single cisterns of rough endoplasmic reticulum. The cytoplasm contains sheaves of fine filaments, 75 Å in diameter. Uranyl acetate and lead citrate, ×17,500.
Electron micrograph showing tenuous cellular processes extending into extracellular vacuoles and desmosomes (arrow) between coapted cell membranes. Uranyl acetate and lead citrate, ×10,000. Right: Electron micrograph showing an island of cytoplasm resulting from invagination by a cytoplasmic process from an adjacent cell. The island is bounded by double membranes with pinocytotic vesicles. Uranyl acetate and lead citrate, ×22,000.

Extracellular vacuoles resulting from irregular dilatation of the intercellular space are a common feature in ecchordoses and chordoma. Due to the complex interdigitations of the encompassing cells, some of these vacuoles may be surrounded by cytoplasm and appear intracellular. However, they can be readily distinguished from the true intracytoplasmic vacuoles by the microvilli and pinocytotic vesicles that line the extracellular vacuoles.

Mitochondria encircled by cisterns of rough endoplasmic reticulum are prominent in some of the cells of ecchordosis and have been observed in chordoma and notochord. The dumbbell-shaped mitochondria described by Erlandson, et al., were not observed in these ecchordoses and have not been demonstrated in other ultrastructural studies of chordoma.

Except for the presence of mitotic figures in chordoma, as illustrated by Murad and Murthy, ecchordosis and chordoma are morphologically quite similar. No ultrastructural features accounting for the difference in biological behavior were observed in this study.

References
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Received for publication June 10, 1970.
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