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Case Reports

Rhabdomyosarcoma of the Oesophagus

Light and Electron Microscopic Study of a Rare Tumor

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Summary. A 61-year-old man was operated for a large tumor, 12 × 4 cm in size, in the lower third of the oesophagus. The tumor had the appearance of a pleomorphic rhabdomyosarcoma showing cross striations by light microscopy and typical sarcomeres by electron microscopy. This is the fifth undisputed oesophageal rhabdomyosarcoma described in the literature and the first examined using electron microscope.

Key words: Oesophagus – Rhabdomyosarcoma – Electron microscopy.

Introduction

Twelve primary rhabdomyosarcomas of the oesophagus have so far, to our knowledge, been described in the literature. The diagnostic cross striations have been demonstrated in only four of these (Wolfensberger, 1894; Stout and Lattes, 1957; Sumiyoshi et al., 1972; Wobbes et al., 1975). We wish to report a fifth undisputed case. It also demonstrates the usefulness of electron microscopy in the precise diagnosis of rhabdomyosarcoma, especially in cases in which light microscopy fails to demonstrate the typical cross-striated elements of rhabdomyoblasts.

Material and Methods

1. Case Report. A 61-year-old man with severe stenocardia and diabetes mellitus had suffered from epigastric pain and vomiting for one year. He had lost 25 kg of weight and was cachectic at admittance.

On oesophagogastroscopy a large tumor in the lower third of the oesophagus was found. Biopsy showed malignancy. After 14 days of hyperalimentation a right thoracolaparotomy was performed. The tumor was 12 × 4 cm in size and almost obstructed the lower oesophagus. The

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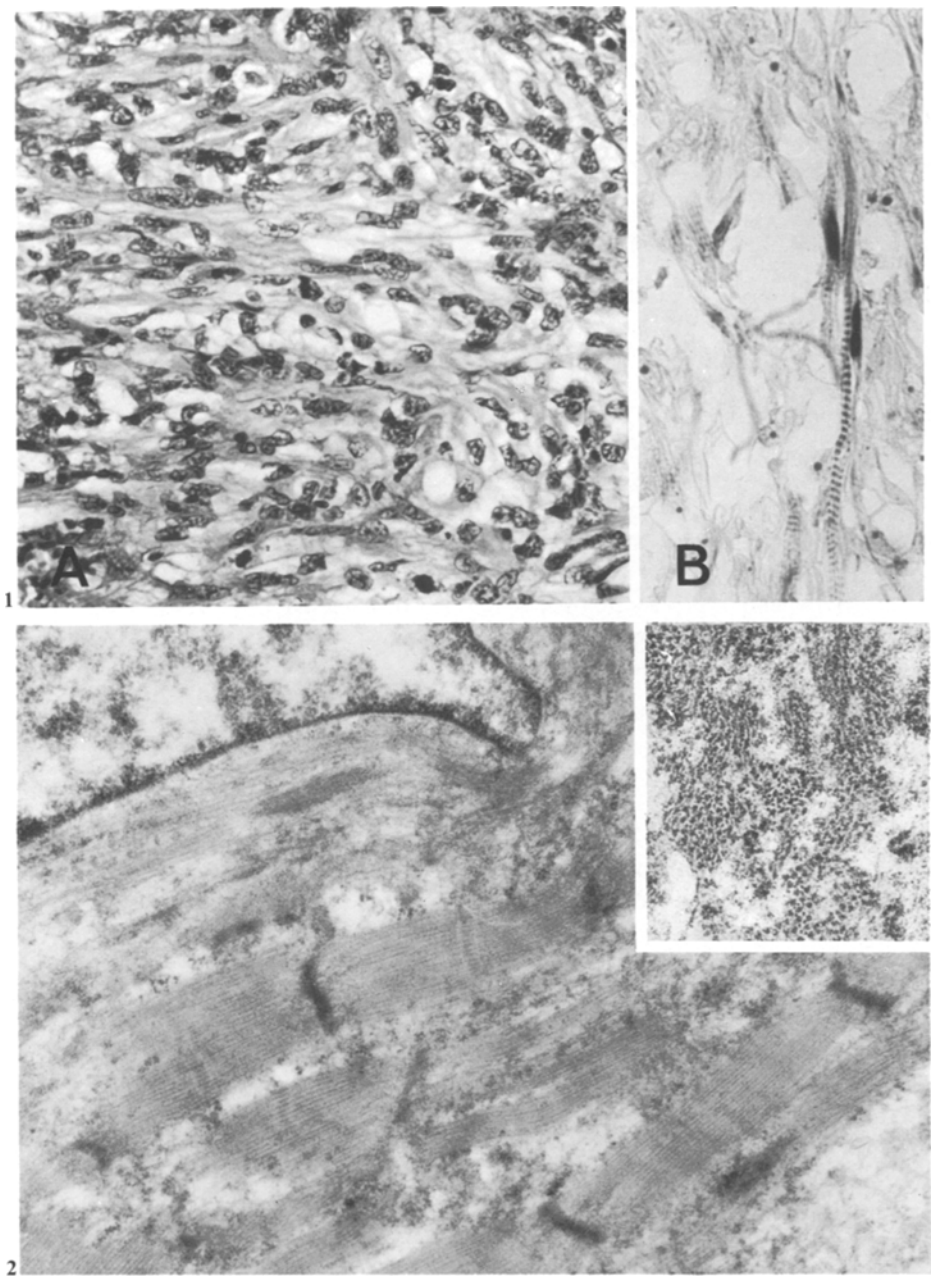

Fig. 1. A. Pleomorphic rhabdomyosarcoma with spindle-shaped cells, having atypical nuclei and finely fibrillar cytoplasm. Haematoxylin and eosin. ×280. **B.** Clear cross striations can be seen in the cytoplasm of several cells. PTAH-staining. ×450

Fig. 2. Electron micrograph of a highly differentiated rhabdomyoblast showing sarcomere structures with Z-bands. ×22,200. Inset shows actin and myosin filaments in cross section with hexagonal arrangement. ×44,800

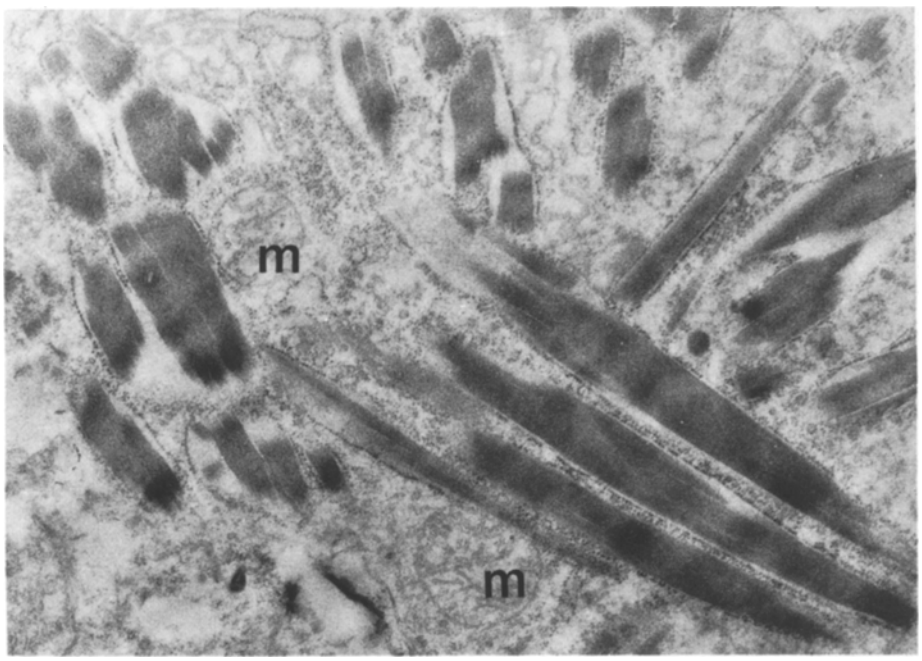

Fig. 3. Intracytoplasmic collagen-like material surrounded by rough endoplasmic reticulum; *m*, mitochondria. ×16,500

thoracic oesophagus and the regional lymph nodes together with the gastric cardia were resected. Continuity was restored by oesophagogastrostomy. Pyloromyotomy was performed. Despite uneventful anesthesia, surgery and immediate postoperative period the patient suffered a severe myocardial infarction on the tenth postoperative day. One month after surgery the patient died from a second myocardial infarction in a general hospital. Autopsy was not performed.

2. Techniques. Paraffin-embedded tumor material was stained with van Gieson, haematoxylin and eosin, and phosphotungstic acid haematoxylin (PTAH) for light microscopical examination. For electron microscopy the formalin-fixed material was postfixed in 1% phosphate-buffered (pH 7.2) osmium tetroxide, and embedded in epoxy resin.

Results

Macroscopically the tumor was connected with the mucous membrane of the oesophagus by an area of ulceration. Metastatic deposits were seen in lymph nodes.

Light microscopically the epithelium of the oesophagus was normal in thickness or, in some areas, thinner than normal. Some superficial ulcerations were seen. In a few sites there was definite cellular atypia with several mitoses and disturbances in polarity corresponding to severe dysplasia. The tumor was highly cellular consisting of mainly elongated, spindle shaped and highly polymorphic nuclei with one or more prominent nucleoli (Fig. 1 A). The cytoplasm was slightly basophilic and fibrillar. Many mitoses were seen. In PTAH-staining many trans-

verse striated elements were present (Fig. 1 B). The histological picture was consistent with pleomorphic rhabdomyosarcoma.

Electron microscopically the nuclei were large with deep indentations and contained one or more prominent nucleoli. A moderate amount of mitochondria, glycogen and free ribosomes were seen in the cytoplasm. Two types of intracytoplasmic filaments were present: thin (actin) and thick (myosin). In some cells a sarcomeric pattern with Z-bands could be seen (Fig. 2). Usually the filaments were distributed in a random fashion. Both longitudinal and transverse sections were seen in the same plane. In cross sections a typical hexagonal arrangement was present (Fig. 2). In one cell dense intracytoplasmic material surrounded by rough endoplasmic reticulum was seen (Fig. 3). No clear banding of this material could be verified; however, it resembled the collagen-like intracytoplasmic material described in some human sarcomas (Levine et al., 1978). Some cells contained tight junctions in the cytoplasmic membrane. A basal lamina surrounded some cells.

Discussion

According to Goodner et al. (1963), 0.5% of all malignant oesophageal tumors are sarcomas, fibrosarcoma being the most common form. Including the present case, twelve reports of rhabdomyosarcoma of the oesophagus have so far, to our knowledge, been reported in the literature. Some of these form the sarcomatous element of carcinosarcoma (Stout and Lattes, 1957).

The tumors occur most frequently in the central and distal parts of the oesophagus (Wobbes et al., 1975). Of the reported patients, aged from 27 to 78 years, nine have been male and three female. According to Ewing's classification, the oesophageal sarcomas are subdivided into ulcerative and polypoid forms. The former metastasize rapidly, infiltrate deeply and cause loss of blood. In the present case the tumor was ulcerated and infiltrative. Metastases were found in the draining cardiac lymph nodes.

It has been proposed that the rhabdomyosarcomas of the oesophagus originate from undifferentiated mesenchyme which, during embryonal evolution becomes localized in the foregut (Templeton and Heslin, 1961). This hypothesis is useful particularly when rhabdomyosarcoma is localized distally in the oesophagus (DeMuth, 1956), since there is no striated muscle tissue in this region of the organ. In the central portion of the oesophagus there is both striated and smooth muscle tissue, while proximally there is only striated muscle (Bloom and Fawcett, 1975).

The diagnosis of rhabdomyosarcoma by light microscopy is difficult because the tumor usually consists of undifferentiated cells. The demonstration of cross-striations in these cells by light microscopy has succeeded only in five oesophageal rhabdomyosarcomas, including the present case (Wolfensberger, 1894; Stout and Lattes, 1957; Sumiyoshi et al., 1972; Wobbes et al., 1975). There are also opinions that the presence of transverse striations is not essential for the diagnosis (Horn and Enterline, 1958).

Electron microscopically, however, the diagnosis can be made easily even from routine formalin-fixed material by showing actin (60 to 80 Å) and myosin (120 to 150 Å) filaments. Especially typical is the hexagonal arrangement of these filaments in cross sections. The finding of whole sarcomeres with Z-bands, as in the present case, and of a sarcotubular system is not essential for the diagnosis.

The intracytoplasmic collagen-like material seen in the present case has been described in osteogenic sarcomas, liposarcomas, malignant fibrous histiocytomas and rhabdomyosarcomas (Levine et al., 1978). The material is usually free or inside a smooth membrane, and it has been suggested that it is a product of degradative activity within tumor cells (Levine et al., 1978). Since the material in our case seems to be inside rough endoplasmic reticulum, a possible disturbance in the synthetic process has also to be considered. The presence of intracellular collagen in these tumors emphasizes a common histogenesis from a primitive mesenchymal cell.

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