Histol Histopath (1993) 8: 593-598

Histology and Histopathology

Primary papillary psammomatous adenocarcinoma of the umbilicus

N. Hernández, V. Medina, H. Alvarez-Arguelles, R. Gutiérrez, J. Pérez-Palma and L. Díaz-Flores
Departments of Anatomy, Pathology and Surgery, Faculty of Medicine, University of La Laguna, Canary Islands, Spain

Summary. The histological and ultrastructural features, as well as the immunoreactivity of one case of uncommon primary papillary and psammomatous adenocarcinoma of the umbilicus are studied in the present work. The observations have been undertaken in a nine-year follow-up, and have included the primitive tumour, two local recidives, and inguinal lymphatic metastasis on two occasions. Papillary structures, numerous psammoma bodies, as well as weak and focal positive reactions to CEA and cytokeratin were present in all the tumours. Since these features and their ultrastructural characteristics were identical to primary papillary serous neoplasias of the peritoneum and ovarium, the hypothesis of an origin in coelomic remnants is considered.

Key words: Psammomatous adenocarcinoma, Umbilicus, Ultrastructure, Immunohistochemistry

Introduction

The primary papillary carcinoma of the umbilicus rich in psammoma bodies is an extremely rare neoplasm (Steck and Helwig, 1965a; Glazer, 1973; Ross and Hill, 1975), which must be differentiated from secondary carcinomas (Steck and Helwig, 1965a; Barrow, 1966; Samitz, 1975).

In the absence of a location other than the umbilicus, its origin has been related to umbilical embrionary endodermic remnants (Jauniaux et al., 1989), principally the urachus (Steck and Helwig, 1965b; Ross and Hill, 1975) and the omphalic mesenteric duct (Moore, 1956; Steck and Helwig, 1964; Glazer, 1973). Nevertheless, there are numerous differences between this type of tumour and those originating in vitelo-intestinal and urachal components.

Given these considerations, the present work was undertaken to study the pathological features of the primary papillary carcinoma of the umbilicus and to compare their light microscopic, ultrastructural and immunohistochemical characteristics with those described for papillary serous peritoneal tumours (Raju et al., 1989). Our findings provide evidence that the coelomic mesodermic remnants of the umbilical duct are the most likely origin of this neoplasm.

Offprint requests to: Dr. N. Hernández, Departamento de Cirugía, Facultad de Medicina, Universidad de La Laguna, Islas Canarias, Spain

Materials and methods

Clinical presentation

A 62-year-old woman, who in 1982 underwent surgery for a small umbilical indurated tumour, which, according to the patient, was noticed 32 years before, becoming enlarged and violet-coloured shortly before the operation. The histological study showed the presence of a papillar carcinoma. All the complementary tests: thyroid scintigraphy; hepatic scintigraphy; barium enema; upper gastrointestinal X-ray series; intravenous pyelogram; abdominal CT; and exploratory laparotomy, showed the lack of any primary neoplasm in any other localization.

One year later, the patient returned to the hospital because of an umbilical recidive. After removal of the lesion, the diagnosis of papillar and psammomatous carcinoma was confirmed. New complementary tests showed an absence of different available sources of the primary tumour other than the umbilicus.

Four years later, the patient presented right inguinal enlarged lymph nodes that were removed, showing metastases of a papillar and psammomatous carcinoma. New complementary studies and a new laparatomy did not show any different primary focus aside from the previous umbilical one.

One year later, the patient returned, this time with bilateral inguinal enlarged lymph nodes; the histological examination showed the same pattern seen in the previous studies.

After this operation and until the present moment, the patient remains without recurrence.

Histological procedures

Tissue for light microscopy was formalin-fixed. Several paraffin sections were prepared and stained with haematoxylin and eosin, Mallory trichrome, as well as with alcian blue (pH 2.5) and periodic acid-Schiff (with and without hyaluronidase and diastase pretreatment). Polyclonal carcinoembryonic antigen (CEA) and cytokeratin (K 506 and K 518 Dako PAP Kits, respectively) immunoperoxidase stain were performed using the avidin-biotin immunoperoxidase technique.

Results

The microscopic characteristics were similar in the original tumour to those in the two local recidives and in the inguinal lymphatic metastases. All the tumours were papillary (Fig. 1) with some areas of solid growth pattern and dense connective tracts. The papillary structures were well formed, with fibrovascular stalks and an epithelial lining of columnar or cuboidal cells (Fig. 2), sometimes pseudostratified and with tuft formation. The proliferating epithelial cells showed a polygonal morphology, with eosinophilic cytoplasms and oval crowded nuclei. Mitoses were relatively numerous (Fig. 2). A higher nuclear cytoplasmic ratio, with irregularity and overlapping of the nuclei was observed in areas of confluent epithelial growth. The cells were occasionally epithelial mucin-positive, but the reaction was poor. Numerous psammoma bodies were present (Fig. 1). These calcific concretions with concentric basophilic laminations showed different sizes. The smaller ones were among tumoral cells, as well as in the fibrovascular cores or in the interpapillary stroma (Fig. 3). When joining, these spherules became calcified masses or aggregates (Fig. 3). Reactions to CEA and cytokeratin were positive, but weak and focal.

Ultrastructurally, the above-referred to aspects were confirmed in the epithelial cells. The nuclei, with prominent nucleoli, and rich in euchromatin, were oval and indented (Fig. 4). The cytoplasms contained some cisternae of rough endoplasmic reticulum, oval mitochondria, variable numbers of vesicles, pleomorphic lysosomes, poorly developed Golgi apparatus and a few filaments. Short, narrow and scarcely branched microvilli were observed in the apical surface (Fig. 5). Between epithelial cells there were well-formed desmosomes, gap junctions, some plasma-membrane interdigitations and terminal bar complexes (Fig. 5). The latter being at the apices of the cells. A basal membrane was observed separating the epithelial cells from the papillary stalks. Occasionally, focal intracytoplasmic condensations, tumour cell necrosis, or thickening of basal lamina followed by calcification were present (Fig. 6).

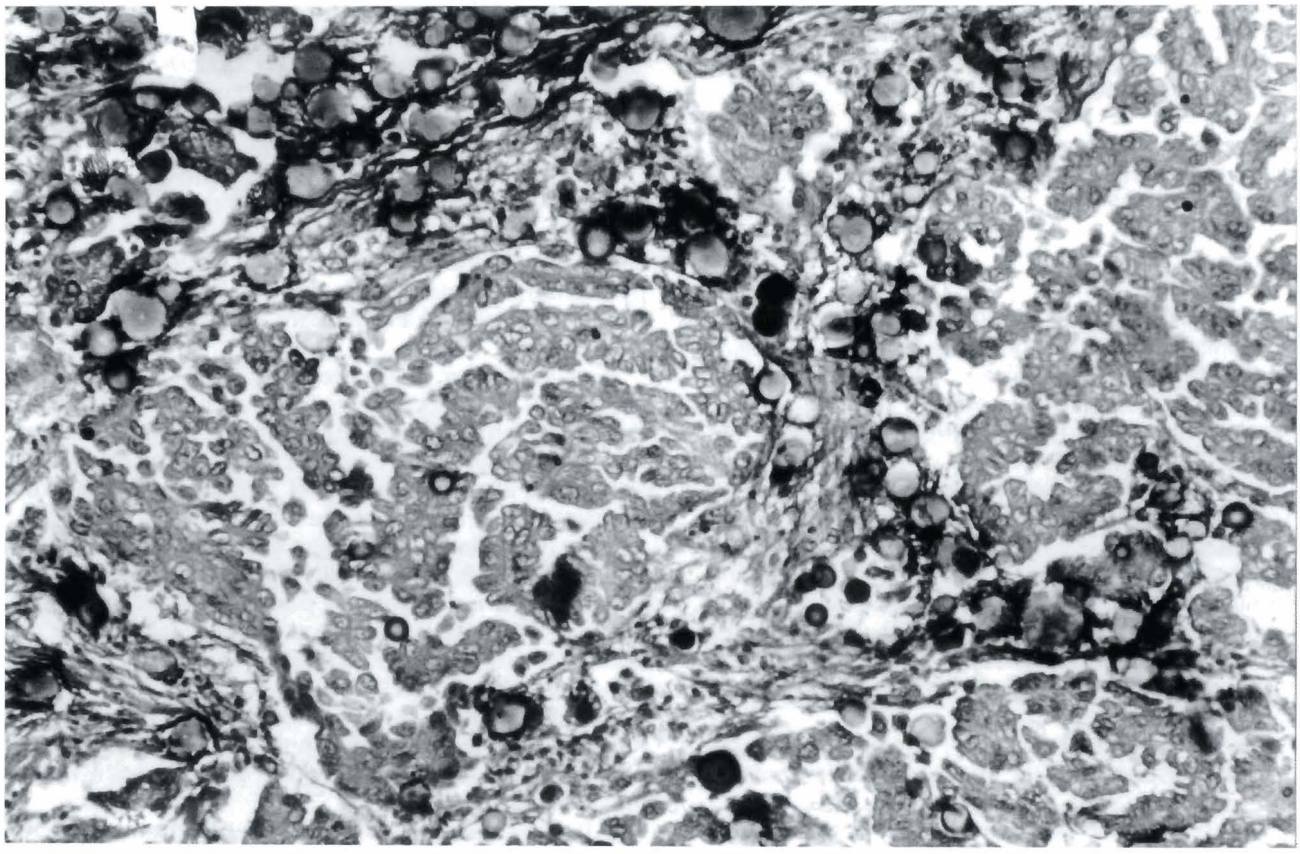

Fig. 1. Papillary carcinoma of the umbilicus with numerous psammoma bodies. Columnar or cuboidal cells lining the fibrovascular stalks are observed. Haematoxylin-eosin stain. x 90

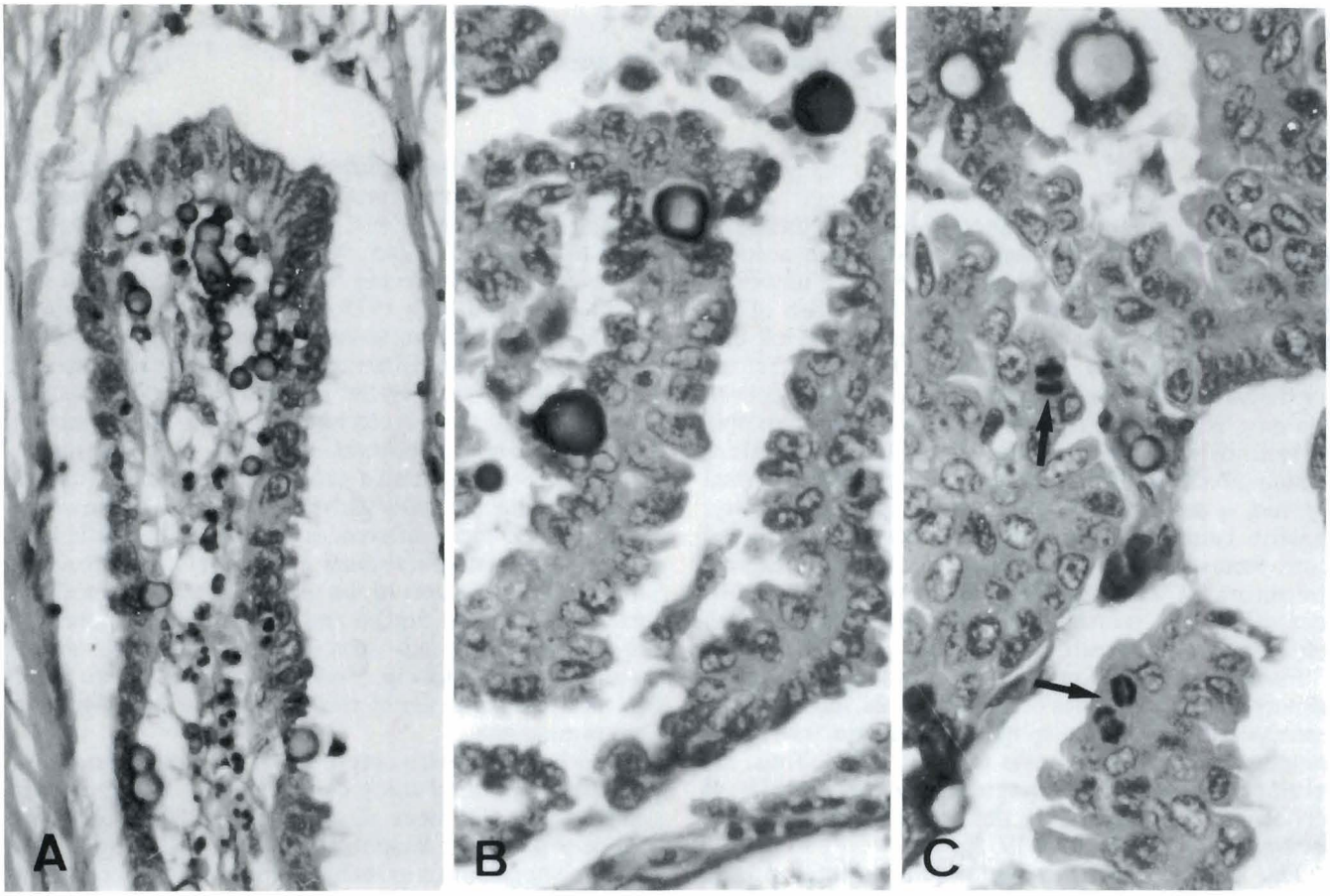

Fig. 2. Appearance of papillae in papillary carcinoma of the umbilicus. **A and B)** Delicate papillae with fibrovascular stalks and psammoma bodies. The latter are observed among tumoral cells and in fibrovascular cores. **C)** Several mitosis are observed (arrows). Haematoxylin-eosin stain. x 160

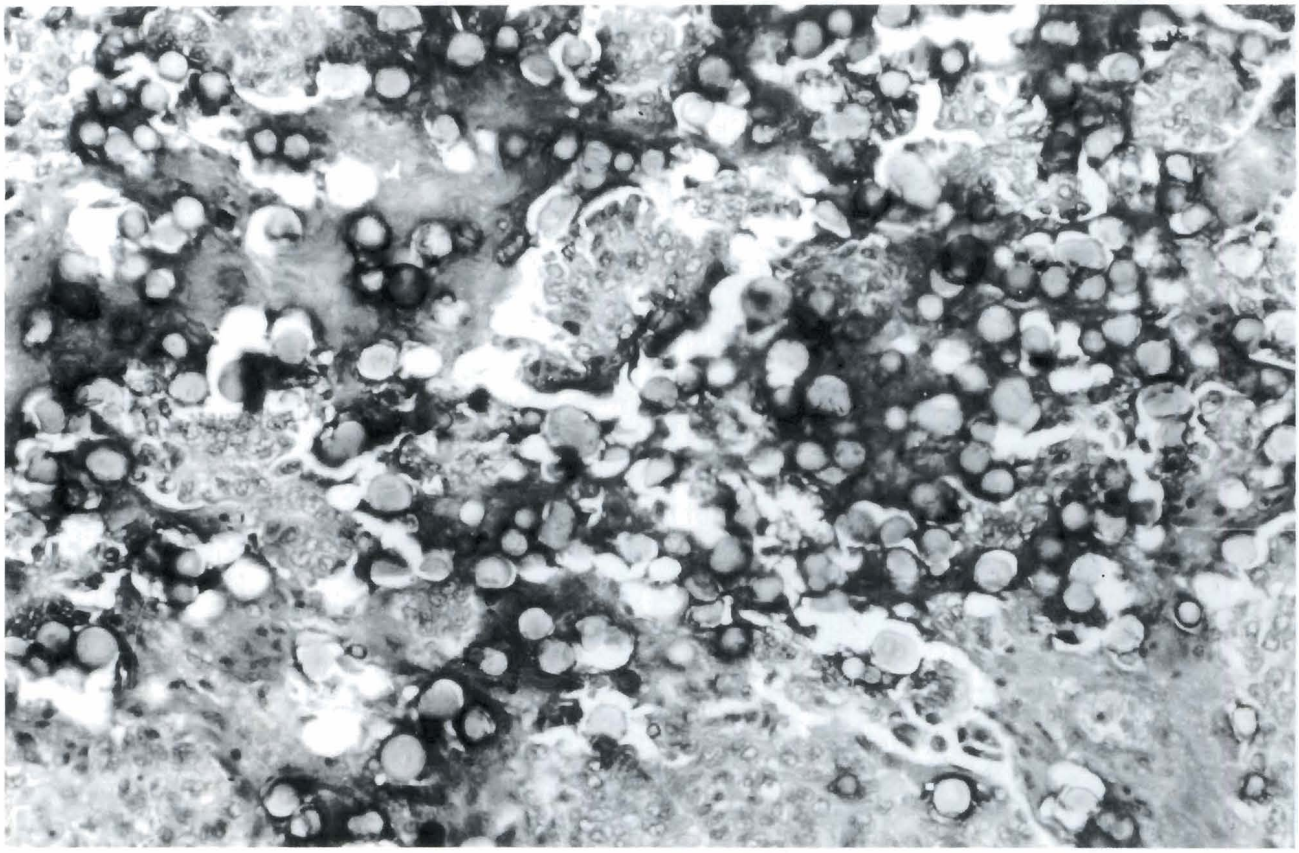

Fig. 3. Psammoma bodies are observed in the interpapillary stroma making up aggregates. Haematoxylin-eosin stain. A) x 90, B) x 120

Discussion

Umbilical carcinomas are almost always secondary to metastasis from visceral tumours. Taking into account the published statistics in this field, primary umbilical malignant neoplasms represent only 1/4 or 1/6 of the metastatic tumours at the same location (Steck and Helwig, 1964; Barrow, 1966; Glazer, 1973). Therefore, our diagnosis of primary umbilical malignant neoplasm was only accepted by default, excluding other possible extraumbilical neoplasms by means of clinical examination and complementary tests, including: normal thyroid scintigraphy; normal hepatic scintigraphy; negative barium enema; negative upper gastrointestinal X-ray series; negative intravenous pyelogram; negative abdominal CT; and two laparotomies without demonstration of primary ovary neoplasm. This has been confirmed after ten years of follow-up.

Furthermore, among the few reported types of primary adenocarcinomas of the umbilicus, most of them reproduce intestinal-like epithelial structures formed by columnar mucosecretory cells (Glazer, 1973). Thus, the primary papillary and psammomatous umbilical adenocarcinoma type, as in the present case, is a very rare tumour (Ross and Hill, 1975).

The origin of primary adenocarcinomas of the umbilicus has been explained from tissular remnants in this place. The comparative study of the vitelo-intestinal (omphalo-mesenteric) duct and the urachus (Trimingham and Mc Donald, 1945; Moore, 1956; O'Leary and O'Leary, 1964; Steck and Helwig, 1964, 1965b; Glazer, 1973; Ross and Hill, 1975; Jauniaux et al., 1989), with the histological characteristics of the neoplasms, which reproduce intestinal-like epithelium, suggests that the latter could arise from the omphalo-mesenteric duct of the umbilicus. On the contrary, the origin of the primary papillary adenocarcinomas presents great difficulty, having been considered a neoplasm originating from the umbilical urachal sinus (Ross and Hill, 1975). However, not only are there differences of the tumoral cells with those of the vitelo-intestinal components, but also with those of the remnants of the umbilical urachal sinus. For example, both the uracho and its remnants are lined by urothelium, frequently surrounded by a smooth muscle wall (Steck and Helwig, 1964). These facts are not found in the reported cases of this tumoral variant, including the present one.

Recently, primary sero-papillar adenocarcinomas of the peritoneum, histologically identical to the ovarian ones, have also been described (McCaughey, 1980; Foyle et al., 1981; Shapiro and Nunez, 1983; Raju et al., 1989), suggesting a possible coelomic origin for these

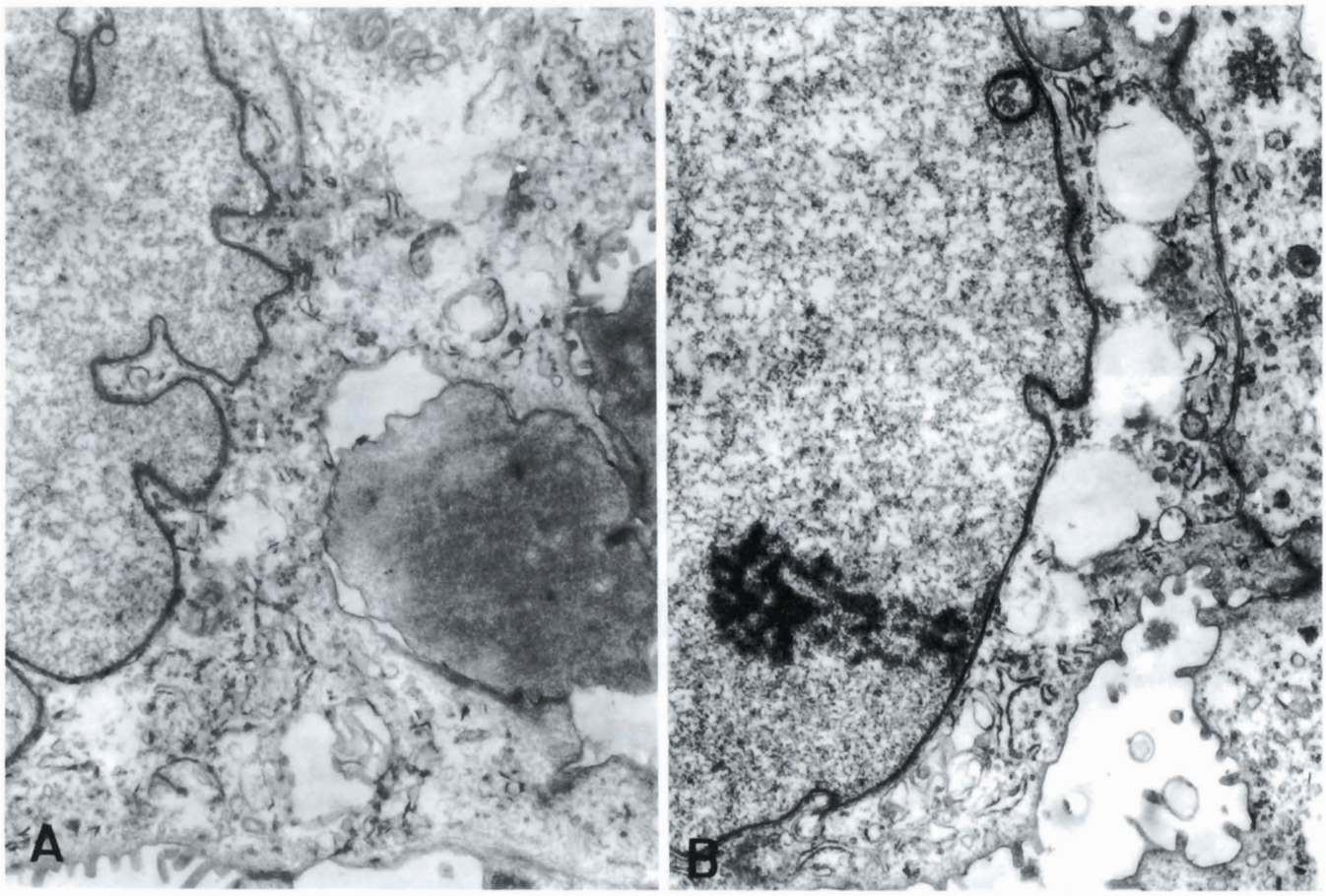

Fig. 4. Ultrastructure of epithelial cells. The nuclei are indented and rich in euchromatin, showing prominent nucleoli. x 14,000

tumours. Since, according to our observations, the light microscopic characteristics of the above mentioned tumours are also identical to those of the primary papillary psammomatous umbilical adenocarcinoma, the hypothesis of an origin in coelomic remnants could be extended to the latter. In fact, during the embrionary period, the coelom invaginates the initial portion of the umbilical cord, conforming a space named omphalic or umbilical coelom, in which the intestinal loops are initially situated, constituting the so-called physiological umbilical hernia. At the end of the third month of embryological development, the umbilical coelom disappears. There is the possibility that from these coelomic remnants of the umbilical duct, papillary neoplasms, similar to the ovarian ones, could develop.

Our immunohistochemical and ultrastructural observations reinforce this idea, given the similar aspect found between the present case and some types of ovarian and peritoneal papillar and psammomatous tumours (Taylor, 1988; Raju et al., 1989). All of them could, therefore, have the same origin in the primitive coelom. In our opinion, the endodermic origin from urachus remnants (Ross and Hill, 1975) is less likely.

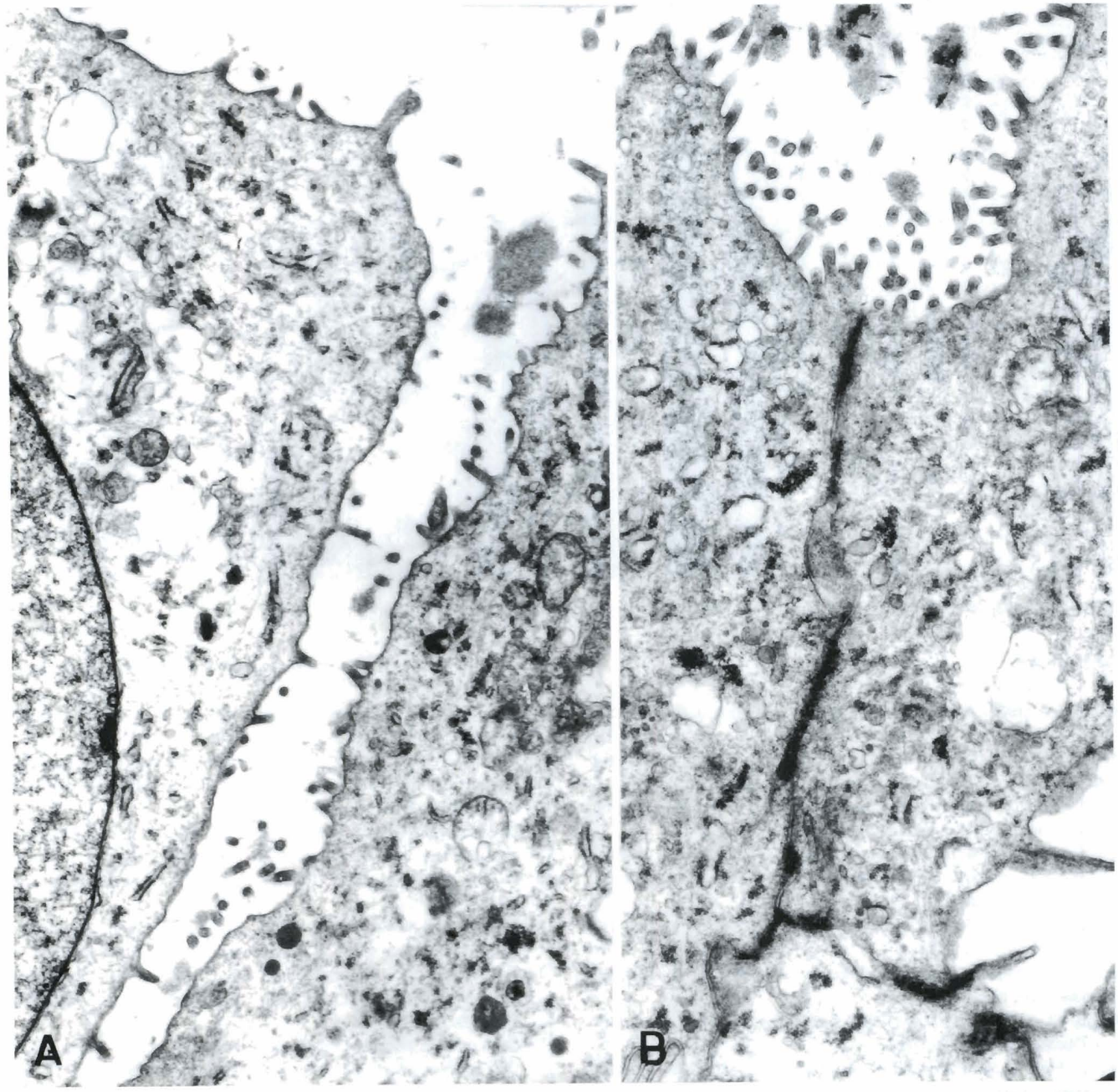

Fig. 5. Short, narrow and scarcely branched microvilli are observed in the apical surface of the epithelial cells **(A)**. In **B** intercellular junctions with desmosomes and gap junctions are shown. x 16,000

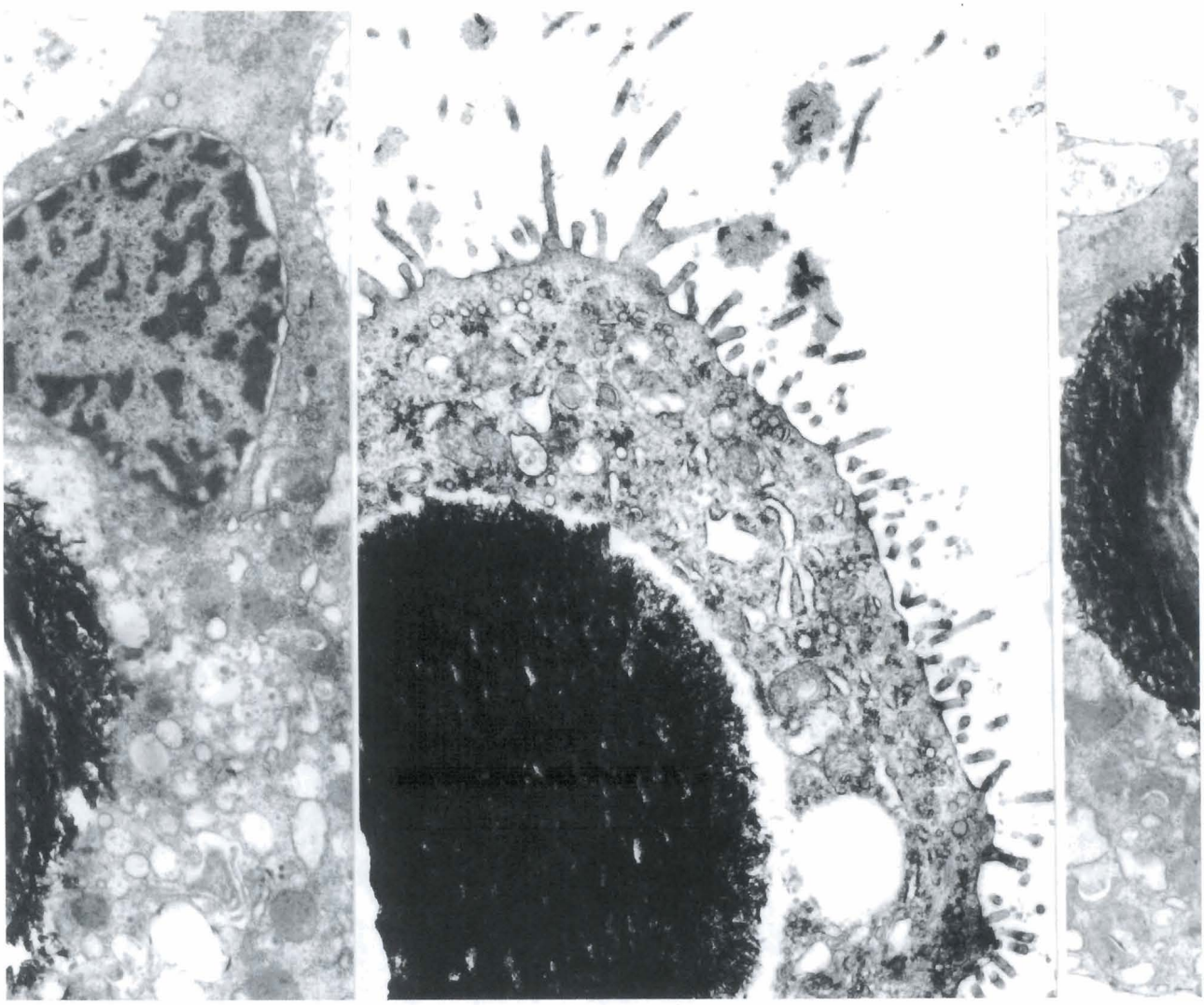

Fig. 6. Ultrastructural images of typical calcified psammoma bodies. x 16,000

References

Barrow M.V. (1966). Metastatic tumors of the umbilicus. J. Chronic Dis. 19, 1113-1117.

Foyle A., Al-Jabi M. and McCaughey W.T.E. (1981). Papillary peritoneal tumors in women. Am. J. Surg. Pathol. 5, 241-249.

Glazer G. (1973). Primary adenocarcinoma arising in a vitello-intestinal duct remnant at the umbilicus. Br. J. Surg. 60, 247-249.

Jauniaux E., De Munter C., Vanesse M., Wilkin P. and Hustin J. (1989). Embryonic remnants of the umbilical cord: Morphologic and clinical aspects. Hum. Pathol. 20, 458-462.

McCaughey W.E. (1980). Papillary peritoneal neoplasms in females. Pathol. Annu. 20, 387-404.

Moore T.C. (1956). Onphalomesenteric duct anomalies. Surg. Gynecol. Obstet. 103, 569-580.

O'Leary J.L. and O'Leary J.A. (1964). Carcinoma of the umbilicus. Am. J. Obstet. Gynecol. 89, 136-137.

Raju U., Fine G., Greenawald K.A. and Ohoródnik J.M. (1989). Primary papillary serous neoplasia of the peritoneum. A clinicopathologic and ultrastructural study of eight cases. Hum. Pathol. 20, 426-436.

Ross J.E. and Hill R.B. (1975). Primary umbilical adenocarcinoma. Arch. Pathol. 99, 327-329.

Samitz M.H. (1975). Umbilical metastasis from carcinoma of the stomach. Arch. Dermatol. 111, 1478-1479.

Shapiro S.P. and Nunez C. (1983). Psammoma bodies in the cervicovaginal smear in association with a papillary tumor of the peritoneum. Obstet. Gynecol. 61, 130-134.

Steck W.D. and Helwig E.B. (1964). Cutaneous remnants of the onphalomesenteric duct. Arch. Dermatol. 90, 463-470.

Steck W.D. and Helwig E.B. (1965a). Tumors of the umbilicus. Cancer 18, 907-915.

Steck W.D. and Helwig E.B. (1965b). Umbilical granulomas, pilonidal disease and the urachus. Surg. Gynecol. Obstet. 120, 1043-1057.

Taylor C.R. (1988). Immunomicroscopy. A diagnostic tool for the surgical pathologist. Vol. 19. Major problems pathology. Saunders Co. W.B. Philadelphia.

Trimingham H.L. and Mc Donald J.R. (1945). Congenital anomalies in the region of the umbilicus. Surg. Gynecol. Obstet. 80, 152-163.

Accepted April 5, 1993