Acta Dermatovener (Stockholm) 51: 460–466, 1971

SYRINGEAL HIDRADENOMA—AN UNUSUAL ECCRINE TUMOUR

Edward Shmunes, Allan Izumi and Herman Beerman

From the Department of Dermatology, University of Pennsylvania School of Medicine, Philadelphia, Pennsylvania, USA

Abstract. An unusual organoid tumour of the scalp of eccrine derivation is reported. The histopathological, histochemical, and ultrastructural characteristics of the tumour all tend to confirm this derivation. The tumour appeared locally invasive but its ultimate malignant potential remains uncertain. Based on the histologic features, the process has been designated *syringeal hidradenoma.* No report of a similar lesion could be uncovered in the literature.

Tumours of the scalp can be classified as either primary or secondary. Primary adnexal tumours of the scalp are uncommon. Metastatic tumours to the scalp usually arise in breast, gastrointestinal tract, lung, genitourinary tract and pelvic organs, and are rare (2). In most instances, differentiation of metastatic tumour from primary adnexal tumours is not difficult. On rare occasions, metastatic renal cell carcinoma or adenocarcinoma may pose a histopathologic problem in differentiation (4). The following case report describes a unique eccrine tumour of the scalp which occurred in a patient with a lung malignancy.

CASE REPORT

A 50-year-old Caucasian physician noted a persistent, minimally pruritic, erythematous, crusted area on his scalp which allegedly followed trauma in that region 2 years prior to hospital admission. The past medical history included adult onset of diabetes mellitus and kidney stones.

An 8.5 cm by 5.5 cm, erythematous to violaceous, flat, sclerotic tumour with indistinct margins and telangiectasia was located in an area of male pattern alopecia on the anterior mid-portion of the scalp (Fig. 1). Within, and in adjacent areas of the scalp, several serosanguinous crusted lesions were noted. There was no clinical evidence of hyperhidrosis in the tumour site. There was no adenopathy in the retroauricular, posterior or anterior cervical lymph nodes.

With the exception of moderate obesity, posterior subcapsular lenticular opacities, and hyperpigmented, slightly depressed scars on the extensor surfaces of the legs, the remainder of the physical examination revealed no abnormalities.

A number of laboratory studies gave normal results: (total leukocytes and differential count, hemoglobin, hematocrit, platelet count, clotting time, prothrombin time, prothrombin consumption, clot retraction, recalcification time, Factor V, urinalysis, Rapid Plasma Reagin Test, blood urea nitrogen, serum sodium, potassium, chloride, carbon dioxide content, triglycerides, cholesterol, lactic dehydrogenase, alkaline phosphatase, uric acid, calcium, phosphate). The serum glutamic oxalacetic transaminase was 50 units, serum glutamic pyruvate transaminase 48 units, serum creatinine 1.1 mg%, total protein 8.2 g%, albumin 3.6 g%, alpha$_1$ globulin 0.3 g%, alpha$_2$ globulin 0.7 g%, beta globulin 1.2 g%, gamma globulin 1.3 g%.

Intermediate strength purified tuberculin protein derivative skin test as well as coccidiodin skin test were negative at 24 and 48 hours. Roentgenographic examinations of the skull were interpreted as normal and failed to show any detectable bone or soft tissue changes in the area of the scalp tumour. Roentgenographic examination of the chest showed an irregular 3 cm nodular density in the right base. The electrocardiogram was interpreted as possibly reflecting old inferior myocardial infarction. An electroencephalogram was interpreted as within normal limits.

A right lung middle and upper lobectomy and peritracheal lymph node dissection were performed. The pathologic diagnosis of the lung tumour was "adenosquamous carcinoma of the lung". Tumour was found in 1 of 7 peribronchial lymph nodes.

MATERIALS AND METHODS

Four biopsy specimens from four different areas of the scalp lesion were obtained. Portions of tissue were fixed in formalin and stained with hematoxylin and eosin, PAS, and PAS diastase. Fresh specimens were also processed for the following enzymes: amylophosphorylase (1), succinic dehydrogenase (10), indoxyl esterase (5), leucine aminopeptidase (1), acid phosphatase (6), alkaline phosphatase (5), glucose-6-phosphate dehydrogenase (3), triphosphopyridine-nucleotide diaphorase (12), and ATP-ase (11).

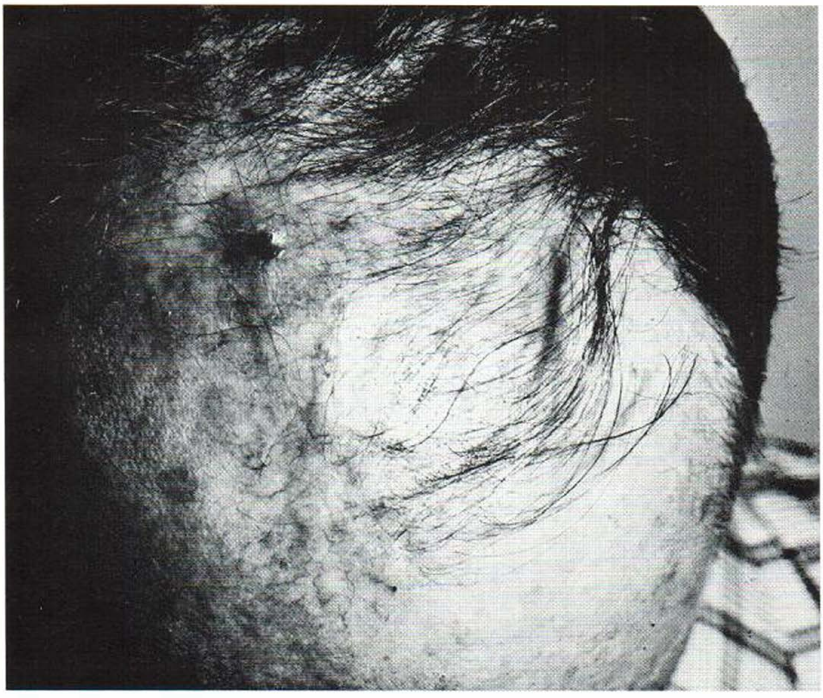

Fig. 1. Large lesion in mid-portion of anterior scalp. Note ill-defined borders and telangiectasia. Serosanguinous crust is present.

Another portion of tissue was immediately cut into small pieces, fixed in a cold 10% glutaraldehyde solution, then in a 1.0% osmic acid solution in sodium phosphate buffer adjusted to pH 7.3. After one hour's fixation, the pieces of tissue were dehydrated, embedded in Araldite and prepared for electron microscopy.

RESULTS

Histopathology

Punch biopsies obtained from a crusted area and from the anterior portion of the sclerotic lesion on May 4, 1970, revealed a tumour in the dermis composed of cell islands and tubules, mostly two cells thick surrounded by a fibrous stroma. Some of the tubules were dilated and contained casts. The process went to the lowermost portion of the specimen. No sweat glands were present but there was an occasional hair. On May 13, 1970, a deeper specimen was obtained. This specimen showed features similar to those in the punch biopsy specimens but there were numerous sweat glands, sebaceous glands, hair follicles, and thick-walled blood vessels as well as dilated capillaries. The tubules were even present in the fat (Figs. 2 and 3). Nowhere in the specimen was there definitive evidence of epithelioma nor did the picture resemble the malignant pulmonary tumour this patient had. All the features suggested that the tumour was of eccrine origin. Serial section of all tissue revealed the uniform picture described above. The histological picture was interpreted, for want of a better designation, as syringoma, although it was fully realized that the clinical features were not at all in keeping with the usual type of lesions.

The specimens were studied by Dr Hermann Pinkus who hesitated to identify the tumour with the process ordinarily known by the name syringocystadenoma multiplex. He believed that this was an extraordinary tumour which was related to eccrine tissue, and which looked benign in its upper portion, but which became much less mature in the deeper portions and resembled basal cell epithelioma. We adopt Dr Pinkus' proposal of the name syringeal hidradenoma for this tumour.

Histochemistry

Sections studied by a variety of procedures and evaluated on a 3+ scale by Dr Bernard Czernobilsky are as follows:

	Cells
Acid phosphatase	+
Alkaline phosphatase	0
Esterase	+

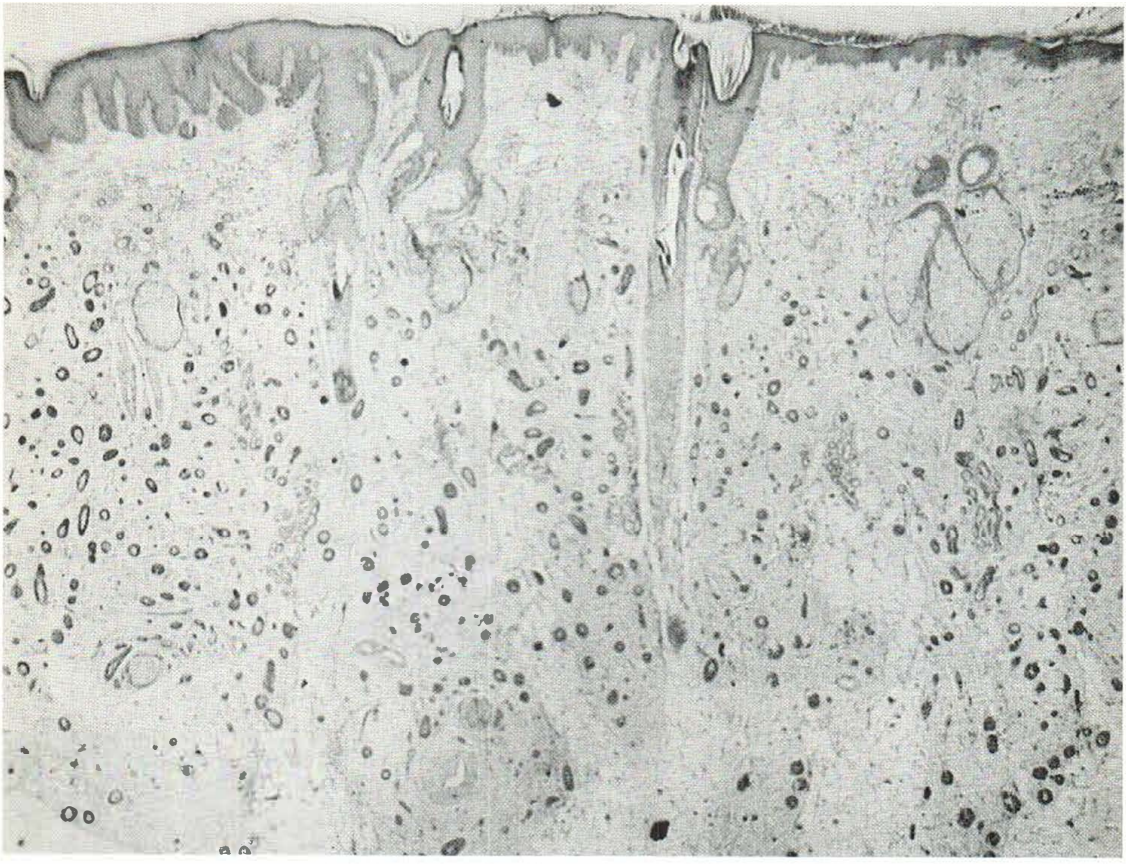

Fig. 2. Closely aggregated cystic or tubular structures, surrounded by a fibrous stroma. Tumour extends into subcutaneous tissue. × 23.

ATP-ase	±
Succinic dehydrogenase	+ +
Glucose-6-phosphate dehydrogenase	±
Triphosphopyridine nucleotide	± to +
Leucine aminopeptidase	0
Phosphorylase	+ +

Electron microscopy

The ultrastructural features of this tumour resembled those features which have been described as characteristic of immature intra-epidermal eccrine sweat duct (8). Luminal cells showed a periluminal tonafilamentous zone, well developed microvilli, prominent desmosomes, and abundant ribonucleoprotein particles. However, few multivesiculated bodies were noted, and numerous mitochondria were observed (Figs. 4 and 5).

DISCUSSION

The clinical features of this tumour were characterized by its flat quality, sclerosis, telangiectasia, erythema, and poorly delineated margins. The crusted lesions within and adjacent to the tumour were excoriated and healed during hospitalization.

The histopathologic features of this tumour closely resembled those of syringoma. Several aspects of the histopathologic changes were atypical: (1) the duct-like structures were more closely aggregated and separated by less connective tissue stroma, (2) portions of the tumour extended below the subcutaneous tissue, (3) in these subcutaneous extensions the tumour changed its configuration to a more solid type of cord or nest.

The occurrence of a strong reaction for both phosphorylase and succinic dehydrogenase, as well

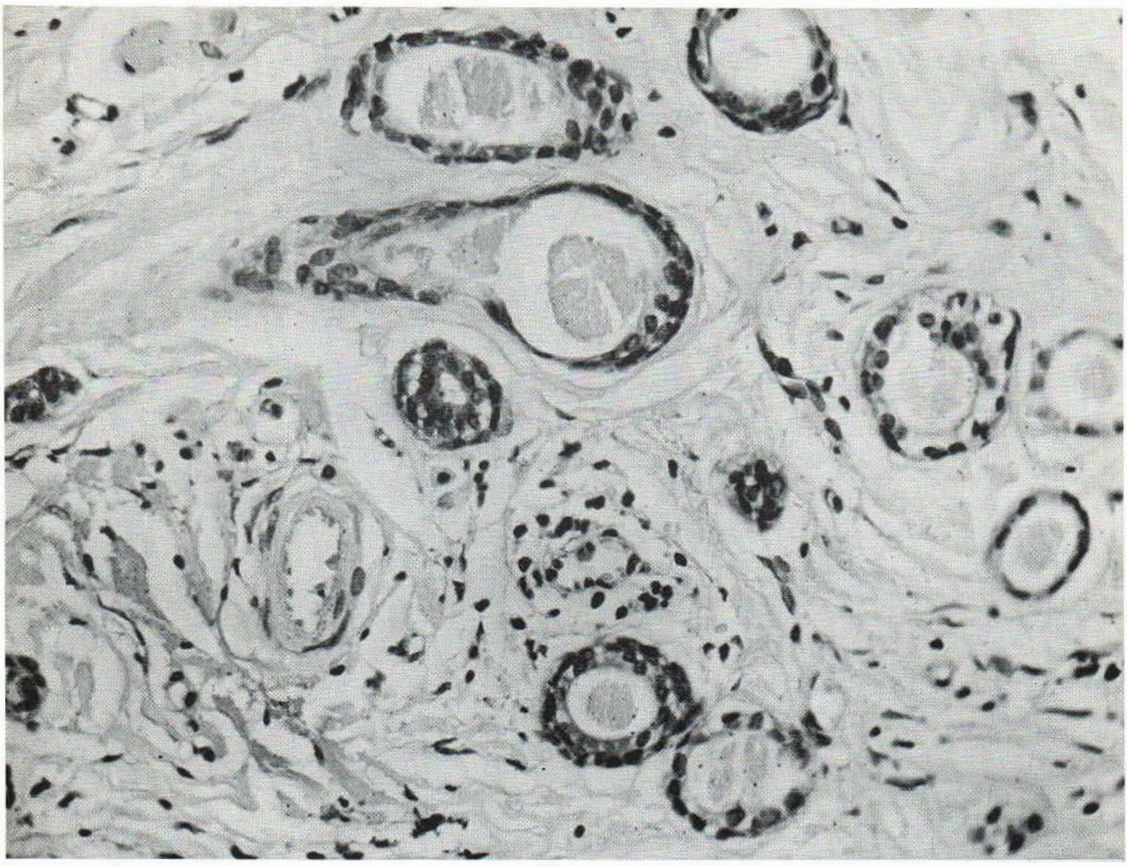

Fig. 3. Higher magnifications of dilated tubular structures. Note syringoma-like pattern. × 340.

as the presence of diastase resistant PAS-positive material within the tubules indicated that the tumour was of eccrine origin. Furthermore, sections stained for acid phosphatase, characteristically strongly positive in apocrine structures, were weakly positive in this tumour. The weak staining reaction for alkaline phosphatase indicated that the tumour was probably of ductal or acrosyringeal origin (9).

The ultrastructural features of embryonic eccrine intra-epidermal duct have been defined as the presence in the luminal cells of multivesiculated bodies (lysosomes), keratohyaline granules, and a periluminal tonafilamentous zone. This combination of elements in one cell has been used as criteria for differentiation in the direction of eccrine intraepidermal duct, and eruptive hidradenoma and syringoma have been found to possess these features.

The tumour in our patient shows well developed microvilli, cyst formation, and a periluminal tonafilamentous zone. Multivesiculated bodies were few and mitochondria were numerous.

Based on the clinical and histopathologic evidence, eccrine nevus, poroma, spiradoma, and adenocarcinoma were removed from consideration.

It is important to distinguish this tumour from basal cell epithelioma with eccrine differentiation described by Freeman & Winkelmann (7). In their first case, the original clinical presentation was in 1926 at which time it was described as a 2 to 3 cm firm linear lesion at the vertex of the scalp of a 34-year-old white woman. It was excised, electrodessicated, then re-excised 4 and 6 years later, respectively. Recurrences were noted subsequently on four more occasions, requiring repeated excision and skin grafting. Although a clinical picture

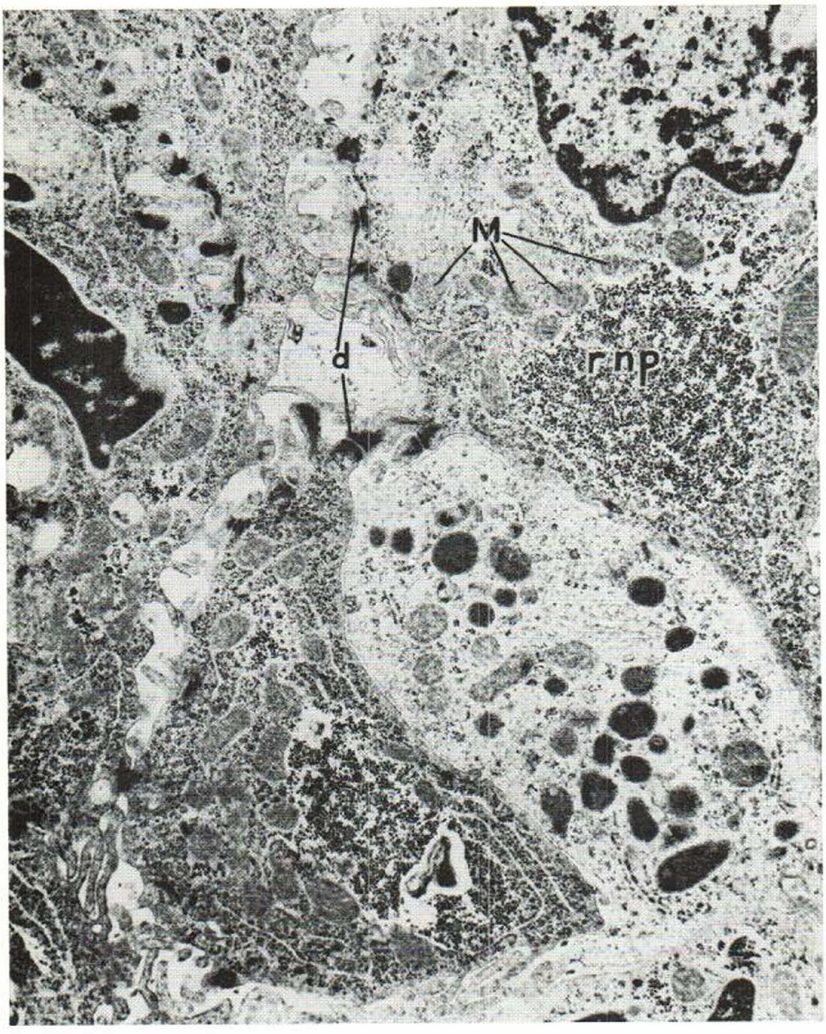

Fig. 4. Area of adjoining tumour cells showing prominent desmosomes (*d*) and abundant ribonucleoprotein (*rnp*) and mitochondria (*M*). × 19 100.

of the tumour taken in 1966 bears some resemblance to the tumour in our patient, the repeated instrumentation undoubtedly influenced its appearance.

In their second case, the tumour was described as a moveable cutaneous mass with a raised smooth pearly border surrounding a crusted ulcer, occurring on the neck of a 49-year-old white male. This latter description of a moveable tumour bears little clinical resemblance to our patient's tumour.

The histopathological descriptions of the tumours reported by Freeman & Winkelmann were not homogeneous. In the first case they describe islands of basaloid cells, islands of cells with alveolar spaces within them, cell masses with multiple lumina, and a stroma that included fibrous and myxomatous areas. In their second case, there were different appearances in several areas of the tumour, including basal-like, cystic, syringoma-like, and sclerosing-like features.

In contradistinction, the histopathologic changes in our tumour show a homogenous picture of syringoma-like and cystic structures throughout the entire dermis except at the subcutaneous level where tumour extensions show solid nests and cord-like arrangements.

Electron microscopic study of the cystic areas of the tumours described by Freeman & Winkelmann "revealed a space containing a few amorphous electron-dense particles bordered by a thin layer of the typical polygonal tumor cells but

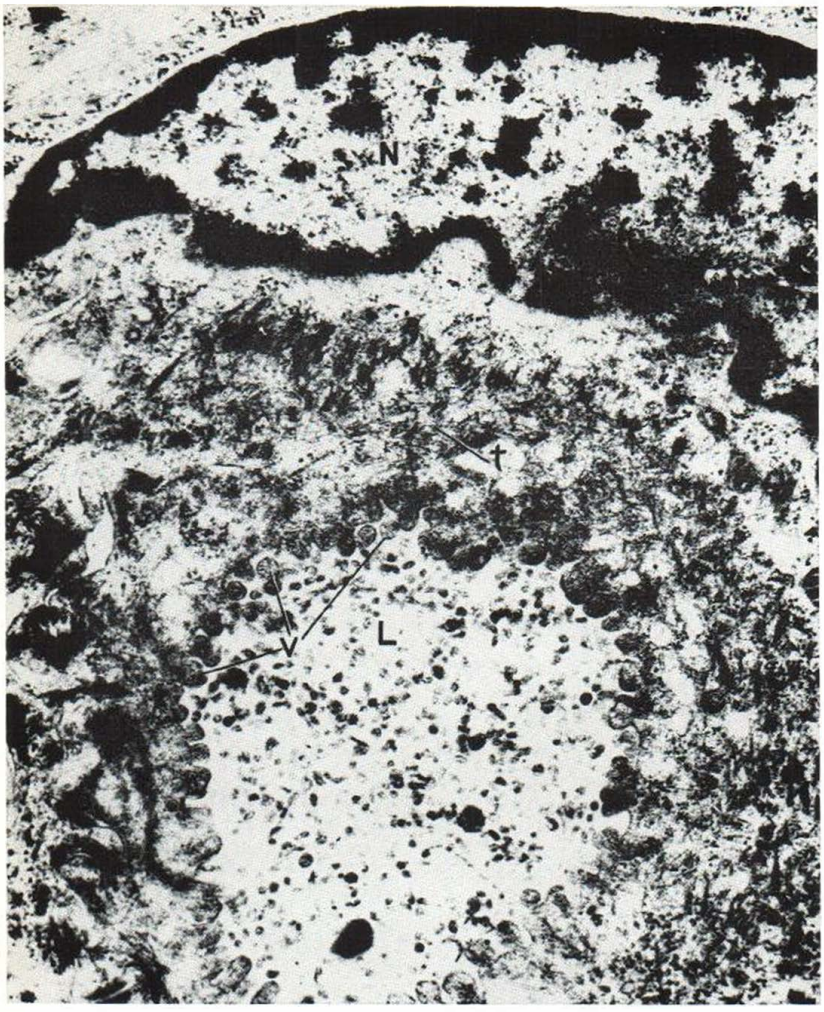

Fig. 5. Luminal cell with nucleus (*N*) showing numerous microvilli (*v*) extending into lumen (*l*), and perilumi-nal tonafilamentous zone (*t*). × 34 000.

without villi on the luminal border". Ultrastructural study of the cystic areas of our tumour clearly show well-developed microvilli of peri-luminal cells (Fig. 5).

Although similar enzyme activity were reported by Freeman & Winkelmann in their patients, the clinical, histopathological, and ultrastructural differences, define the uniqueness of the tumour in our patient. The only feature in common is that their tumours and ours are apparently all of eccrine origin or differentiation.

ACKNOWLEDGEMENTS

Donald M. Pillsbury, M.D., suggested study of his private patient.

Hermann Pinkus, M.D., interpreted histopathological sections of the tumour. Bernard Czernobilsky, M.D., interpreted the histochemical staining reactions and provided the services of his technician, Miss Jackie Hendricks.

K. C. Tsou, Ph.D., interpreted the electron microscopy and provided the services of his electron microscopy laboratory.

Mr Ed Gilfort provided the photographic services.

REFERENCES

1. Barka, T. & Anderson, P.: Histochemistry: Theory, Practice, and Bibliography. Moeber Medical Division, Harper & Row, New York, 1965.
2. Beerman, H.: Some aspects of cutaneous malignancy: Ruben Nomland Memorial Lecture. Arch Derm (Chicago) *99:* 617, 1969.
3. Cohen, S. & Way, S.: Histochemical demonstration of pentose shunt activity in smears from the uterine cervix. Brit Med J *1:* 88, 1966.

4. Conner, D. H., Taylor, H. B. & Helwig, E. B.: Cutaneous metastasis of renal cell carcinoma. Arch Path *76:* 339, 1963.
5. Czernobilsky, B. & Tsou, K. C.: Adenocarcinoma, adenomas and polyps of the colon: histochemical study. Cancer *21:* 165, 1968.
6. Evans, C., Whinney, C. & Tsou, K. C.: A new histochemical method for acid phosphatase by the use of 5-iodoindoxyl phosphate. J Histochem Cytochem *14:* 171, 1966.
7. Freeman, R. G. & Winkelmann, R. K.: Basal cell tumor with eccrine differentiation (eccrine epithelioma). Arch Derm (Chicago) *100:* 234, 1969.
8. Hashimoto, K., DiBella, R., Borsuk, G. & Lever, W. F.: Eruptive hidradenoma and syringoma: histological, histochemical, and electron microscopic studies. Arch Derm (Chicago) *96:* 500, 1967.
9. Hashimoto, K., Gross, B. G. & Lever, W. F.: Syringoma: histochemical and electron microscopic studies. J Invest Derm *46:* 150, 1966.
10. Pearse, A. G.: Histochemistry: Theoretical and Applied. Little Brown & Co., Boston, 1960.
11. Wachstein, M. & Meisel, E.: Histochemistry of hepatic phosphatases at a physiologic pH: with special reference to the demonstration of bile canaliculi. Amer J Clin Path *27:* 13, 1957.
12. Wattenberg, L. W.: A histochemical study of five oxidative enzymes in carcinoma of the large intestine in man. Amer J Path *35:* 113, 1966.

Received April 14, 1971

Edward Shmunes, M.D.
Department of Dermatology
Hospital of the University of Pennsylvania
Duhring Laboratories Bldg.
Philadelphia, Pennsylvania 19104
USA