922

PATHOLOGISTS' CLUB
OF NEW YORK

WMP (BEH CASE)

MEETING

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Date: **Thursday, October 13, 1994**
(Please note date change for this meeting only from the 1st to the 2nd Thursday of the month)

Place: **Cabrini Medical Center**
227 East 19th Street
(at Third Avenue)
New York, New York 10003

Host: **Dr. Maria T. Sabatini**

Information: **Joan Kelly**
(212) 995-6107

RECEPTION AND DINNER: **5:15 - 7:00 P.M.**
SCIENTIFIC SESSION: **7:00 - 9:00 P.M.**

Please Note: So that we can plan dinner, let us know of your intention to attend by October 6.

Directions: The hospital is located at 227 East 19th Street between Second and Third Avenues.

By Subway: 4, 5, or 6 train to 14th St./Union Square - L (Crosstown) Train to Third Avenue/14th Street. Walk uptown to 19th Street.

By Bus: Second and Third Avenue buses stop at 19th Street.

Parking: Some parking will be available on a "first come" basis on 19th Street East of the main entrance and on 20th Street near the Emergency Room.

Next Meeting: **Bellevue, November 3, 1994**

CASE HISTORIES - PATHOLOGISTS' CLUB MEETING, CABRINI MEDICAL CENTER, 10/13/94

Case # 1 S94-1206 Invited discussant: Dr. Susan Kornacki, Bellevue Hospital
Host discussant: Dr. Maria T. Sabatini

A 34 year old Asian woman was admitted to Cabrini Medical Center with complaints of intermittent epigastric pain and nausea for the past several months. She denied any significant past medical or surgical history. A gall bladder x-ray series at another institution was unremarkable. Upper GI demonstrated an irregularity in the upper portion of the stomach. Upper endoscopy revealed no abnormalities of the mucosa of the esophagus, stomach, or duodenum. CT of the abdomen showed a 4.6 x 2.9 cm mass in the left upper quadrant believed to have arisen from the region of the left adrenal gland. At surgery, the inferior and superior aspects of the mass were attached by filamentous adhesions to the superior pole of the left adrenal and the posterior wall of the stomach respectively. Neither of these organs, however, appeared to be a site of origin for the mass. The specimen was excised, leaving most of the adrenal intact.

Case # 2 S94 2395 Invited discussant: Dr. Abdelmonem Elhosseiny, N Y Medical College
Host discussant: Dr. Gerald Minkowitz

The patient is a 32 year old man with a mass in his neck for many years which gradually increased to its current 10 cm size. Rarely the mass was associated with tenderness; the patient is uncertain whether the mass drained into the mouth. Clinically, the mass is soft, nonmobile, and in the midline. It visibly distends the floor of the mouth. No lymph nodes are palpable. The patient, who is otherwise in good health, smokes and drinks. A fine needle aspirate was performed. The slide submitted consists of a cell block preparation.

Case # 3 A92-019 Invited discussant: Dr. Jaishree Jagidar, Bellevue Hospital, NYU
Host discussant: Dr. Maria T. Sabatini

The patient is a 60 year old white homosexual man, HIV positive for 2 yrs, with a CD-4 count of 194/c.mm, and a history of peptic ulcer disease treated with antacids. There was no past history of opportunistic infections. In December 1991, the patient developed abdominal discomfort and diarrhea. Travel history over the last 20 years included Southeast Asia, the Mediterranean, So. America, and the US Southwest. He was admitted to the hospital because of rapid deterioration and dehydration. A CBC and chemistries showed a macrocytic anemia , HGB 11.6gm/dl, WBC 7000 with 3% eos, and mildly elevated LFTs. Stools were negative for O & P. The patient had GI bleeding and hemorrhagic bronchial secretions. He developed respiratory distress, acute renal failure, and cardiorespiratory failure, expiring on the 11th hospital day. An autopsy was performed.

Case #4 S94-4514 Invited discussant: Dr. Howard Mizrachi, NYU Medical Center
Host discussant: Dr. Ciril-Christian Rizk

A 68 year old white female, with a past medical history of TAH/BSO for an ovarian tumor in 1988 and a right lower lobe resection for undifferentiated carcinoma of the lung in 1993, was admitted to Cabrini Medical Center for emergency small bowel resection due to intussusception. At surgery, a polypoid mass of necrotic red tissue was found protruding into the lumen.

Case #5 S94-2807 Invited discussant: Dr. John Scholes, NYU Medical Center
Host discussant: Dr. Ciril-Christian Rizk

A 44 year old Filipino male with a past history of pneumonia in September 1993 presented in April 1994 with swelling of the right supraclavicular area. At the time of the previous admission, no organisms had been isolated. The patient was treated with intravenous cephalosporins, and his pneumonia slowly resolved. The patient moved from the Philippines 2 years ago and lives in New York. He reports a one month visit to California.

PATHOLOGISTS' CLUB MINUTES CABRINI MEDICAL CENTER
October 13, 1994

CASE #5

The first case discussed was Case #5, that of a 44 year old Filipino male with a history of pneumonia in September 1993 and a travel history to California, who presented in April 1994 with swelling of the right supraclavicular area. The section submitted for evaluation was a full thickness skin biopsy including subcutaneous tissue. Low power revealed a diffuse inflammatory lesion with a vaguely nodular pattern. In the mid-dermis there was perivascular and adnexal involvement. The inflammation was mixed including polys and epithelioid histiocytes. Microabscesses collared by macrophages and an outer zone of lymphoplasmacytic infiltrate were particularly evident in the subcutaneous tissue. An additional slide provided to the invited discussant showed a more distinctly nodular and granulomatous pattern. Dr. Scholes discussed the differential diagnosis for a suppurative granulomatous dermatitis which includes various infections-e.g. deep fungal, mycobacterial, bacterial, and algal- as well as follicular lesions. The approach to the case included a number of special stains, and careful scruitiny revealed PAS and GMS positive 7-8 micron spores. Among the primary cutaneous deep fungal infections are chromomycosis and sporotrichosis, neither of which were appropriate for this case. Of the secondary deep fungal infections, as ranked by overall incidence, there are blastomycosis, coccydiomycosis, cryptococcosis and histoplasmosis. In this case the diagnostic spherules with endospores of coccidiomycosis were identified.

Dr. Rizk agreed with this diagnosis. The patient had been asymptomatic, and was HIV negative with no risk factors. Coccidiomyocosis, a fungus prevalent in the arid and semiarid soil of the south west, is commonly inhaled with dust particles and most commonly produces an asymptomatic pulmonary disease. Only .1 to .5 percent go on to produce systemic disease, but of these 15 to 20 percent develop cutaneous lesions. Interestingly Filipinos are particularly at risk for systemic dissemination. Additional techniques applied in this case included application of DNA probes and a controlled culture, as presented by Dr. Boyle. An additional finding in this case by Dr. Scholes were macrophages with blue dots raising the question of concomitant leishmaniasis, but both the invited and host discussant reached the conclusion that these were breakdown products of inflammatory cells rather than additional organisms.

DIAGNOSIS: CUTANEOUS COCCIDIOMYCOSIS

Refs: Seminars in Dermatology 12(4): 301-9, 1993 Dec. Coccidiomycosis - Review
Clinical infectious diseases 14 Supl 1:S23-9, 1992 Mar. Lab methods for the diagnosis and confirmation of systemic mycoses (review)
J Amer Acad of dermatology 26(1):79-85, 1992 Jan. Clinicopathologic spectrum of specific cutaneous lesions of disseminated Coccidiomycosis

CASE #1

This is a case of a 34 year old Asian woman, admitted with complaints of intermittent epigastric pain and nausea, who was found to have a 4.6 cm. mass in the left upper quadrant which was attached by filamentous adhesions to the superior pole of the left adrenal and the posterior wall of the stomach. At low power Dr. Kornacki showed that the mass was a cystic structure partially lined by readily identifiable mucosa, submucosa and muscle. At higher power the lining ranged from cuboidal to pseudostratified columnar respiratory type mucosa to gastric type mucosa. Adjacent areas showed inflammation and sheets of polyhedral cells with vacuolated cytoplasm, which by immunohistochemical studies proved to be macrophages. Dr. Kornacki's diagnosis, based on the location of the lesion, was gastric duplication.

Intestinal duplications are cystic or tubular malformations with a well developed smooth muscle layer which are lined by a mucosal membrane and are intimately associated with some part of the GI tract. In assigning a particular subtype, location is more important than the histology. These duplications can be

communicating or noncommunicating. Two important differential diagnoses include Meckel's diverticulum or false diverticula. Ileal duplications are most common and are located on the mesenteric border, while stomach and jejunum are the rarest forms. Approximately 60 percent of these are diagnosed in children less than two years old who may present with a palpable mass, vomiting, weight loss, pain, or failure to thrive. They may also be asymptomatic. Usually gastric duplications are cystic (noncommunicating) and are located on the greater curvature. One proposed theory of pathogenesis is abnormal canalization of a GI tract initially obliterated by an epithelial proliferation. Potential complications include perforation, bleeding, fistula formation, obstruction, rarely carcinoma, and peptic ulceration, which may account for the inflammation and macrophage accumulation in this case. The host discussant, Dr. Sabatini, agreed with this interpretation. Her diagnosis was foregut duplication.

DIAGNOSIS: GASTRIC DUPLICATION (FOREGUT DUPLICATION)

CASE #2

The patient is a 32 year old man with a long standing 10 cm. cervical midline mass. A fine needle aspirate was performed and the slide submitted was the cell block preparation. Included in the findings were squames, some atypical, hairs, abundant pink amorphous material, calcifications, and cholesterol clefts. By immunocytochemistry (invited discussant's lab) the amorphous material was thyroglobulin positive. The differential diagnosis discussed by Dr. Elhosseiny included thyroglossal duct cyst and dermoid cyst. Thyroglossal duct cysts may be found anywhere in the midline from the foramen cecum to the level of the thyroid, but usually these lesions are not so large, and they do not contain keratin or hair follicles. Dermoid cysts, however, may be located on the floor of the mouth, below the platysma. Dermoid cysts of the head and neck constitute 7 percent of all dermoids, and may include epidermal, dermal, or teratoid structures.

Additional clinical history supplied by Dr. Minkowitz included the fact that the patient was a Rikers Island inmate who reported having had the mass for "a long time", possibly from childhood. Complicating clinical features were the possibility that the mass was fluctuant or draining into the mouth. The clinicians' impression was that this mass could be a "plunging ranula". or some form of chronic sialadenitis. The possibility of a dermoid cyst was remote in their differential diagnosis, but confirmed by fine needle aspiration. Dr. Minkowitz reviewed the distiguishing features of hamartoma, choristomas (heterotopias), teratomas, and dermoids, thought to be formed by inclusions of ectodermal elements. He reported that microscopic examination of multiple sections failed to reveal thyroid tissue in the specimen and wondered about the possibility of non-specific staining for thyroglobulin. Dr. Elhosseiny however noted that stains for CEA and calcitonin were negative while repeat thyroglobulin stains were again positive.

DIAGNOSIS: DERMOID CYST

Refs: Dermoid cyst of the floor of the mouth and lateral neck. The Laryngoscope 89:296-305, 1979.
Nomenclature of the Developmental Tumors. Batsakis JG. Annals of Otolaryngology Rhinology and Laryngology 93:98-999,1984.
Dermoid cysts of the floor of the mouth. Oral Surg. 8:1149, 1955.
Dermoid cysts of the floor of the mouth. Br J Oral Surg 3:36,1965

CASE #3

The patient was a 60 year old HIV positive homosexual male with an extensive travel history but no past history of opportunistic infections who developed abdominal discomfort, diarrhea, GI bleeding and hemorrhagic bronchial secretions. The patient's clinical status rapidly deteriorated, and he expired on the 11th hospital day. In beginnning her discussion, Dr. Jagirdar noted that the patient's CD4 count (194/mm3) was not overly low, stools for O and P were negative, and blood examination showed anemia

with only 3 percent eosinophils. Bleeding from both the GI tract and tracheobronchial tree suggested the possibility of a systemic disease. Microscopic examination of the section from the heart showed an elongated structure in a vessel with lilac colored nuclei and a cuticle on the surface. The adjacent myocardium was remarkable only for fibrosis , hypertrophy, and lipofucsin. Dr. Jagirdar interpreted the elongated structure as a filariform larva, and made the diagnosis of strongyloides.

Strongyloides is a parasite which is common in the tropics and subtropical southeastern USA where it is found in soil as a fecal contaminant. The disease may develop in anyone who has ever lived in an endemic area. The incidence in HIV positive and HIV negative individuals is the same. The filariform organism is infective, entering through the skin and traveling through dermal vessels to the lung where the organism is swallowed and matures in the GI tract. The eggs hatch, maturing to the rhabditiform, and in autoinfection, a unique feature of this organism, there is transformation again to the filariform form. Clinically the infection can mimic peptic ulcer disease, and in immunocompromised hosts, hyperinfection may result in widespread organ involvement (heart, lung, GU tract, brain, liver,). The internal structure of the filariform larva is highlighted by phase contrast, and a diff-quik stain highlights the nuclei. As the organism inhabits the crypts, organisms are not shed, and hence cultures may be negative. In hyperinfection, eosinopenia may be present. In cases where cultures for O and P are negative and eosinophilia is not present, serologic testing may be helpful. The differential diagnosis for this filariform larva is hookworm (enterobium vermicularis). Treatment with vermectin shows the greatest promise. Dr. Sabatini concurred with Dr. Jagirdar's diagnosis, and noted that at autopsy there was diffuse and widespread involvement by the disease, but evidence of no other infections.

DIAGNOSIS: STRONGYLOIDES

Ref. Gomles et al. Disseminated strongyloidiasis in AIDS: uncommon but important source AIDS 1991, 5:329-332.
Liu and Weller. Strongyloidiasis and other intestinal nematode infections. Infectious Dis Clin N Am 7:655-682, 1993.
Haque et al. Pathogenesis of human strongyloidiasis: Autopsy and quantitative parasitological analysis. Mod Path 7:276-288,1994.

CASE #4

The patient is a 68 year old white female with a past history of an ovarian tumor and undifferentiated carcinoma of the lung, who was operated on for intussuception. At surgery, a polypoid mass was found protruding into the lumen. Sections show a malignant neoplasm involving all layers of the bowel wall and involving vascular channels. The tumor was composed of sheets of moderately pleomorphic cells with abundant pink cytoplasm and variably prominent nucleoli. Mitotic figures including atypical forms were easily identified. The presence of intracytoplasmic pink globules suggested the possibility of a yolk-sac tumor or hepatoma, but as these were seen only in degenerating areas, it seemed more likely they were altered red blood cells. Architecturally, some areas showed a vaguely nested appearance, while other areas appeared spindled. The differential diagnosis considered by Dr. Mizrachi included epithelioid leiomyosarcoma, metastatic carcinoma, and metastatic melanoma. Of the possible primary sites, the patient had a history of ovarian and lung tumors. Primary sites known for malignant neoplasms with abundant pink cytoplasm include liver, kidney, and adrenal. Of these, Dr. Mizrachi would favor metastatic lung or melanoma as those being statistically the most likely. Immunohistochemistry was performed with the following results: muscle specific actin +, S100 strongly +, HMB-45 +, desmin, cytokeratin 5.2, and alpha fetoprotein, all - . On repeat stains the same pattern of reactivity was obtained with only some differences in intensity. Dr. Mizrachi felt comfortable in eliminating the possibility of metastatic carcinoma; the finding of positive HMB 45, however, was puzzling. This antibody has been reported in non-melanocytic lesions such as angiomyolipoma, and Dr. Mizrachi postulated that since gastrointestinal stromal tumors may show schwannian features which in turn may show melanocytic differentiation, it

might be possible for a GIST to stain with the melanoma marker (HMB-45). His diagnosis: epithelioid leiomyosarcoma with anomalous staining for HMB-45, ? divergent differentiation.

Dr. Rizk agreed with the diagnosis of epithelioid leiomyosarcoma or malignant leiomyoblastoma. Intussuception has been reported in up to 30 percent of gastrointestinal smooth muscle tumors. In this case the mass measured 6 x 4 cm. with hemorrhage and ulceration noted at the tip. Microscopically these tumors are composed of an admixture of fusiform and epithelioid cells with variable pleomorphism and mitotic activity. The criteria for malignancy include 4 to 5 mitoses/HPF, size greater than 6.0 cm., and a predominantly epithelioid histology. Immunostains, with the exception of a negative S100, were the same as the invited discussant, including HMB-45 positivity. Electron microscopy showed subplasmalellal dense bodies, rough endoplasmic reticulum, and free ribosomes, but no thin myofiloments. Additional clinical features of note were that the patient's previous ovarian tumor was a benign fibrothecoma, and that the patient's initial lung resection had in actuality been a metastasis of this tumor, rather than a primary undifferentiated carcinoma. An additional differential diagnosis suggested by a member of the audience included GANT (gastrointestinal autonomic nerve tumor).

DIAGNOSIS: EPITHELIOID LEIOMYOSARCOMA

Ref: Intussucepton and leiomyosarcoma of the GI tract in a pediatric patient. Dig dis and sciences 38(10):1933-7, 1993.
Diagnostic utility of desmin. A study of 584 cases and review of the literature. Am J Clin Path 93(3):3-5-144, 1990.