

PATHOLOGISTS' CLUB OF NEW YORK

100F


COMP (SEM CASES)

PRESIDENT
FRED B. SMITH, M.D.
DEPARTMENT OF PATHOLOGY
ST. VINCENT'S HOSPITAL
155 WEST 11TH STREET
NEW YORK, NY 10011

MEETING

Thursday, May 1, 1997
Our Lady of Mercy Medical Center
600 East 233rd Street
BRONX, NY 10466
Host: Dr. Marian Waxman
718-920-9876

VICE PRESIDENT
JOAN G. JONES, M.D.
ANATOMIC PATHOLOGY
EINSTEIN-WEILER HOSPITAL
1825 EASTCHESTER ROAD
BRONX, NY 10461

SECRETARY-TREASURER
STYLIANOS LOMVARDIS, M.D.
DEPARTMENT OF PATHOLOGY
ST. CLARE'S HOSPITAL
428 WEST 52ND STREET
NEW YORK, NY 10019

Reception and Dinner: 5:15-7:00: Educational Center, Conference room, Main Floor
Scientific Session: 7:00-9:00: " " " "

Subway: #2 train (west side IRT Express) to East 233rd St. and White Plains Rd. Walk west one block on E. 233rd.

Metro North: take a Harlem line Bronx local from Grand Central or 125th St. to Woodlawn Station. Walk east one block on east 233rd.

By car from Manhattan: FDR Drive north to Willis Ave. bridge to Major Deegan Expressway north (I-87). Exit East 233rd Street, go east on 233rd about one mile. Cross above Rail track and Bronx River Parkway. Hospital is on right.

By car from Queens and Brooklyn: Via Triboro or Whitestone Bridge, take Cross Bronx Expressway. Take second exit to Bronx River Parkway north. Exit to East 233rd Street (exit 10). Turn right, then left at corner lights. Hospital on right.

PARKING: Garage across from hospital on 233rd (not free). Street parking on Bronx Boulevard (just east of Bronx River Parkway) and on Webster Avenue (north of Bronx R. Pkwy and south of hospital).

CASE HISTORIES

Case 1. Discussants: Dr. Ibrahim Hitti, Northshore Forest Hills Hospital, Queens; Dr. Umadevi Katta, OLMHC

A 43 y/o woman presented with acute right upper abdominal pain. Abdominal CT showed gallbladder with gallstones, and a large calcified mass was seen to the left from aorta at the level of the lower pole of the kidney. At surgery, mobile mass was found at the junction of aorta and iliac vessels. Cholecystectomy and resection of the mass were performed. The specimen consisted of two whitish-brownish partly calcified tissue fragments 7x3x2 cm in greatest dimensions.

Case 2. Discussants: Dr. Constantine Axiotis, SUNY Health Science Center at Brooklyn, Dr. ^LLaura Vahr

A 73 y/o man presented with low grade fever and increasing shortness of breath. The symptoms did not respond to antibiotics treatment. Routine cultures and AFB smear were negative. A CT scan of the chest revealed multiple bilateral pulmonary nodules. Bronchoscopy was negative. Open lung biopsy was performed.

Case 3. Discussants: Dr. Marius Valsamis, Westchester County Medical Center, Valhalla; Dr. Firozali Panjvani,

A 75 y/o woman presented with a headache, double vision and gait disturbances. CT of the head revealed a "hyperdense pineal mass;" and enlargement of the ventricles. MRI of the brain disclosed a lesion of the third ventricle causing aqueductal obstruction with hydrocephalus. At first surgery a biopsy of the lesion and ventriculo-peritoneal shunt were done. At the second surgery most of the lesion was debulked.

Case 4. Discussants: Dr. Lawrence Alpert, Northern Westchester Hospital Center, Mt. Kisco; Dr. Umadevi Katta,

A 31 y/o woman presented with difficulty of swallowing, shortness of breath and pain in the neck, of two weeks duration. For the last three years she was aware of a neck mass which recently grew in size. There was compression and deviation of the trachea. CT of the neck showed a mass in the left lobe and bilateral shoddy adenopathy. Left thyroid lobectomy and isthmectomy were performed. Grossly the mass measured 4.3x4x 3.2 cm, was firm, solid and cystic, and appeared well circumscribed.

Case 5. Discussants: Dr. Claudio Guerrieri, St. Vincent's Hosp. Medical Center, New York; Dr. Marian Waxman,

A 29 y/o woman, G6 P3033 was admitted for abdominal pain, and was subsequently operated for an ovarian mass at her 14th week of pregnancy. Right salpingo-oophorectomy and partial omentectomy were performed. The specimen consisted of a 16x17x12 cm solid mass with extensive hemorrhage and necrosis. The tube & omentum were normal.

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MINUTES OF MEETING

Our Lady of Mercy Medical Center
1 May 1997

Inclement weather proved no obstacle to the surprisingly large and cheerful number of participants in the second spring meeting. The membership unanimously voted in favor of three new members, Dr. Jinsong Liu of New York University Medical Center, Dr. Charles C. Marboe of Columbia University, and Dr. Geraldine C. Dela Fuente of Nassau County Medical Center. Dr. Marian Waxman moderated the scientific session.

Case 1. 43 year old woman with acute upper abdominal pain underwent removal of gallbladder and of a mobile 7x3x2 cm partly calcified, white brown mass at the junction of the aorta and iliac vessels.

Invited discussant: Dr. Ibrahim Hitti, North Shore Forest Hills Hospital

Host: Dr. Umadevi Katta

At the edge of the extensively calcified tumor with microscopic cysts there is benign tissue consisting of ducts, acini and pancreatic islet cells. Tumor cells are pale with nuclei resembling those of islet cells. None of the tumor cells stain for chromogranin. Therefore, one must consider this as a case of solid and cystic tumor. It is thought that the tumor starts as glandular and because of poor vascular support, pseudocysts form. This is a solid and cystic epithelial tumor of the pancreas. (Two other examples of this tumor were presented in the November 1996 and in the April 1997 meetings). Among the non-duct types of pancreatic tumors are included the acinar cell carcinoma, pancreatoblastoma, and solid-cystic tumor. Here, we find features of islets of Langerhans and of endocrine tumors of the pancreas. Histochemical findings suggest incomplete differentiation in both exocrine and endocrine directions. Dr. Katta found focal staining for keratin, but none for NSE, chromogranin, synaptophysin, and CD 31 and 34. Unlike ductal carcinoma, this tumor will not stain with CEA. Malignant behavior correlates with venous invasion, nuclear anaplasia and prominent necrobiotic change. Typically it occurs in young women.

Dx: Solid and cystic epithelial neoplasm of the pancreas.

1. Scialfani LM et al: The malignant nature of papillary and cystic neoplasms. Cancer 68: 153, 1991
2. Nishihara K et al: Papillary cystic tumors of the pancreas. Cancer 71: 82, 1993
3. Stommer P et al: Solid and cystic pancreatic tumors. Cancer 67: 1635, 1991

Case 2. 73 year old man with low grade fever, progressive dyspnea and multiple pulmonary nodules, not responding to antibiotics. Unrevealing bronchoscopy, bacterial cultures and AFB smears led to open biopsy.

Invited discussant: Dr. Constantine Axiotis, SUNY Health Science Center, Brooklyn
Host: Dr. Marian Waxman

The differential diagnosis of diffuse pulmonary infiltrates includes a variety of conditions, both inflammatory (angiocentric immunoproliferative, necrotizing sarcomatoid granulomatosis, Wegener's) and neoplastic (metastatic tumors, lymphomas). The infiltrate is angiocentric, it spares the elastica and does not obscure the periphery of vessels. Wide areas of necrosis tend to coalesce and extend into the interstitium. Within the infiltrate one sees activated lymphoid cells, plasma cells, occasional atypical cells with prominent nuclei, but no eosinophils. The predominant smaller cells stain with CD 3 (T cells) and the larger ones with CD 20 (B cells). Within Jaffe's concept of angiocentric immunoproliferative lesions, typically localizing in extranodal sites but recently reported in lymph nodes, too, several different syndromes are recognized: lymphocytic vasculitis (possibly the earlier phase of AIL), lymphomatoid granulomatosis (Liebow), nasal angiocentric lymphoma (midline malignant reticulosis), polymorphous reticulosis, and malignant lymphoma. These are angiocentric and angiodestructive, and are graded according to the degree of nuclear atypia and necrosis, with grade III easily recognizable as malignant lymphoma on routine histologic appearance. Epithelioid histiocytes and giant cells are not often seen. T cells are believed to be reactive and the larger, atypical cells are EB virus-infected neoplastic cells, which predominate in grade III. The EB virus occurs in CD 56 (natural killer-like) cells. This is a case of pulmonary angiocentric immunoproliferative lesion, gr II, formerly designated as lymphomatoid granulomatosis. Dr. Waxman presented the CT images with multiple nodules in the lung. Scattered cells staining with L 26 and 45RO are identified as B cells. He agrees with the diagnosis of malignant lymphoma, lymphomatoid granulomatosis type. This condition may be manifest in one of two subtypes: EB virus-infected cells expressing B cell markers in a mixture with more numerous, reactive T cells (T cell-rich B cell lymphoma), or as a T cell lymphoma with no evidence of B cells and of EBV. It may occur in the skin in up to 40% of patients, in the CNS, upper respiratory tract and kidney but rarely in bone marrow and spleen. It is twice as common in men than in women. Following chemotherapy the nodules have resolved and the patient remains asymptomatic. (In our May 1996 meeting Dr. Frizzera discussed T cell rich B cell lymphoma, which may in fact constitute a proportion of cases of lymphomatoid granulomatosis).

Dx: Malignant lymphoma, lymphomatoid granulomatosis type

1. Lipförd EH, Margolick JB et al: Angiocentric immunoproliferative lesions. Blood 72: 1674, 1988
2. Guinee D, Jaffe E et al: Pulmonary lymphomatoid granulomatosis. Am J Surg Path 18: 753, 1994
3. Myers JL, Kurtin PJ et al: Lymphomatoid granulomatosis... Am J Surg Path 19: 1300, 1995
4. Koss MN: Pulmonary lymphoid disorders. Semin Diagn Path 12: 158, 1995
5. Oshimi K et al: Lymphoproliferative disorders of natural killer cells. Int J Hemat 63: 279, 1996
6. Medeiros L et al: Localization of Epstein-Barr viral genomes... Am J Surg Path 16: 439, 1992
7. Koss MN, Zelen EH: Low-grade B-cell lymphoma and lymphomatoid granulomatosis. Pathology (Phila) 4: 125, 1996

Case 3. 75 year old woman with headache, diplopia and gait ataxia was found to have a hyperdense

pineal mass with aqueductal obstruction and hydrocephalus. After biopsy and ventriculoperitoneal shunt, the lesion was debulked.

Invited discussant: Dr. M. Valsamis, Westchester County Medical Center

Host: Dr. F. Panjvani

Dr. Valsamis summarized the presentation as pointing to a lesion affecting cranial nerves and long tracts, with a mass in the third ventricle. Morphologically it belongs in the group of small round cell tumors. Many cells are pigmented, and are arranged in sheets; there is no organoid pattern. Some cells exhibit nucleoli. Among the pigmented CNS tumors one must consider meningioma, neuroblastoma, melanoma and schwannoma. This has none of the features of meningioma and schwannoma, therefore these are easily excluded. S-100 stains some cells. Glial fibrillar basic protein marks astrocytes only. Strong staining with vimentin confirms that the tumor is a melanoma. The surgeon did not remove the tumor and it continued to grow. MRI showed it to have the density of subcutaneous tissue, therefore it had to be either fat or a blood clot. But the first MRI is identical. Apparently free radicals in melanin give this density. Mitoses and prominent nucleoli are evident. Meningeal melanocytoma differs from malignant melanoma in not having prominent necrosis, and only few mitoses.

Dx: Malignant melanoma

1. Barut S: Primary leptomeningeal melanoma. Neurosurg Rev 18: 143, 1995
2. O'Brien TF et al: Meningeal melanocytoma. Arch Pathol Lab Med 119: 542, 1995

Case 4. 31 year old woman with dyspnea, dysphagia and neck pain for two weeks had known of a neck mass which grew larger in the recent past. It compressed and deviated the trachea. The mass was in the left lobe of the thyroid and there was bilateral shotty lymphadenopathy. The resected left thyroid lobe contained a 4.3x4x3.2 cm firm, solid and cystic, well circumscribed mass.

Invited discussant: Dr. L. Alpert, Northern Westchester Hospital Center

Host: Dr. U. Katta

Dr. Alpert found that the nodule is only partially encapsulated and lacks invasion either of vessels or of the capsule. Some portions are obviously of follicular thyroid tissue; the spindle shaped cells do not represent sarcomatous change in benign tissue, and some polyhedral cells show partial clearing. In places there are suggestions of transition from spindle to polyhedral cells. Could this be a mixed medullary follicular carcinoma? The stroma, which is eosinophilic and clearly produced by clear cells, stains with congo red and exhibits apple green birefringence. Thyroglobulin is demonstrable in both colloid and cells, even in the undifferentiated cells of the solid portion. Surprisingly, there is no reaction to calcitonin either in cells or in the amyloid stroma. Medullary carcinoma virtually always stains with CEA, often with calcitonin, synaptophysin, NSE and cytokeratin. There is no CEA staining in this tumor. Among possible alternatives, follicular adenoma or carcinoma with solid spindle cell component is very rare. The follicular variant of medullary carcinoma is excluded because of the thyroglobulin. It lacks the features of medullary carcinoma with entrapped follicles. Can it be mixed medullary and follicular carcinoma? This tumor is best classified as a tumor of uncertain malignant potential.

Dr. Katta noted the transitions from follicular to spindle cells and minimal atypia. Some foci in the spindle cell component show staining with keratin, thyroglobulin and vimentin. Chromogranin staining has been shown to be more sensitive than calcitonin. Her diagnosis is atypical follicular adenoma.

Dx: Atypical follicular adenoma of thyroid

1. Hazard JB, Kenyon r: Atypical adenoma of the thyroid. Arch Path 58: 554, 1954
2. Rosai J et al: Tumors of the thyroid gland. Atlas of Tumor Pathology, 3rd Series. 1992, pp.38-45.
3. Lang W et al: The differentiation of atypical adenomas. Virch ArchivPathAnat 385: 125, 1980

Case 5. In her fourth week of pregnancy, a 29 year old woman, G6P3A3, required sight salpingo-oophorectomy and partial omentectomy for an 18x17x12 cm solid ovarian mass. The tumor showed hemorrhage and necrosis. Tube and omentum were normal.

Invited discussant: Dr. C. Guerrieri, St. Vincent's Hospital

Host: Dr. M. Waxman

Dr. Guerrieri took note of the absence of any detectable pattern in the tumor and of the merging with the non-neoplastic stroma, proving that single tumor cells enter the stroma: these features help exclude epithelial tumors and point to stromal neoplasms. Plump spindle cells with ample cytoplasm lack nuclear atypia and grooves, and show about 3 mitoses per 10 HPF. There are also round cells, typical of luteinized stromal cells, therefore it belongs in the category of thecoma with focal fibrosis. The vascular pattern shows branching, resembling hemangiopericytoma. Reticulin fibers enclose individual cells, helping to exclude diffuse granulosa and Sertoli cell tumors. Thus the evidence indicates a pure gonadal stromal tumor. Necrosis is not a sign of malignancy, but possibly of ischemia. There is staining with vimentin. Actin stains most spindle cells, but not the luteinized ones. Inhibin, a stain for sex cord cells, marks luteinized but not the spindle cells. Therefore this is a thecomatous tumor with luteinization. Among the population of darker cells, some are large and show mitotic activity, others occur in groups with dark nuclei, raising the possibility of extramedullary hemopoiesis. His diagnosis is luteinized thecoma. One third of cases occur in women under 30 years, are unilateral, solid and yellow, and almost all are benign. They are associated with sclerosing peritonitis. The question of behavior of luteinized thecoma hinges on mitotic activity. In the MGH series, benign prognosis was associated with fewer than 5 mitoses per 10 HPF and no nuclear atypia, potentially malignant course with more than 5. The diagnosis here is luteinized cellular thecoma with myeloid metaplasia.

Dr. Waxman pointed to the large areas of necrosis. His diagnosis is luteinized adult granulosa cell tumor with extramedullary hemopoiesis. The Leder stain highlights the myeloid cells but not erythroid. The omentum also contains clusters of hemopoietic elements. Other tumors in which extramedullary hemopoiesis has been found are hepatoma, meningioma, hemangioma and bronchial carcinoid.

Dx: Luteinized adult granulosa cell tumor/thecoma group with extramedullary hemopoiesis

1. Gregorios JB et al: Extramedullary hematopoiesis in a malignant meningioma. Neurosurg 13: 447, 1983
2. Lara JF, Rosen PP: Extramedullary hematopoiesis in a bronchial carcinoid tumor. Arch Path Lab Med 114: 1283, 1990