

Department of Pathology

Mid-Winter Anatomic Pathology Seminar

"Neoplastic and Non-neoplastic Conditions of the Gastrointestinal Tract"

Guest Consultant: Sandy Templeton, M.D.
Department of Pathology
University of Massachusetts School of Medicine

St. Paul Ramsey Hospital Auditorium
Thursday, January 30, 1975
6:00 PM
Buffet will be served.

CASE #1

This 64 year old female was first seen with a chief complaint of a "swelling by the ear". Upon examination there was a large "tumor" of the parotid gland. The tumor together with a portion of surrounding parotid gland was excised; the gross specimen measured 5 x 3.5 x 2 cm. and consisted of moderately firm gray tissue.

The patient had no other complaints and the physical examination was otherwise negative.

Submitted by: Sam Leung, M.D.
Quain and Ramstad Clinic
Bismarck, North Dakota

CASE #2

This 17 year old female was admitted to St. Paul Ramsey Hospital with a chief complaint of tarry stools for one week plus an intolerance of spicy foods for an indefinite period of time. She also had noted mild right upper quadrant pain which was dull and intermittent and was relieved by sitting upright. One of the many examining physicians elicited a history of bloody stools for at least one year. At the time of admission her hemoglobin was 5.5 grams. An extensive workup followed; a summary of the many radiographic and endoscopic examinations was that of a "polypoid mass in the gastric antrum with the differential diagnosis including focal gastric hyperplasia, gastric polyp, gastric carcinoma, lymphoma, and nodular leiomyoma". A laparotomy was performed; a partial gastrectomy plus splenectomy and liver biopsy followed, upon the recommendation of the pathologist. The gastric tumor was located along the greater curvature and consisted of a firm multinodular mass which protruded into the lumen of the stomach but also protruded from the serosal surface. Individual nodules were described as being confluent but several smaller nodules were identified on the serosal surface. The tumor measured 9 cm. in greatest diameter and the cut surface was light yellow to tan and varied in texture from soft and fleshy to moderately firm.

Submitted by: Pathology Staff
St. Paul Ramsey Hospital

CASE #3

This 30 year old male presented with a chief complaint of midline upper abdominal pain for approximately one year. There was no history of nausea, vomiting, diarrhea, or melena. An initial upper GI series showed a "tennis ball sized gastric mass with central ulceration". At laparotomy with partial resection of the stomach there was a 5 cm. in diameter, firm, round gastric tumor which appeared to arise from the muscularis. Its cut surface was generally a gray color but there were areas of yellowish-gray foci and semitranslucent foci. The edges appeared sharply demarcated from the surrounding gastric tissue. There was a 4 mm. ulceration on the gastric surface of the tumor. Numerous lymph nodes showed no abnormalities.

Submitted by: G. J. Obert, M.D.
St. John's Hospital
Fargo, North Dakota

CASE #4

This 70 year old male was referred to St. Paul Ramsey Hospital because of multiple gastric polyps found on upper GI series. The patient complained of abdominal swelling, decreased appetite, constipation, melena and a weight loss of 10 to 12 pounds. An upper GI series done by his local physician revealed multiple polyps of the stomach and a barium enema was normal. Family history was negative.

On admission to the hospital the patient had a serum calcium of 12.6 mg%, phosphorous of 2.6 mg%, total protein of 5.5 grams%, and an albumin of 3.42 grams%. An IVP showed a left renal cyst of 4.5 cm. and this was the same size as his previous IVP's. Upper GI showed multiple gastric polyps and bone and liver scans were normal. Biopsies of the polyps at time of gastroscopy showed only chronic inflammatory changes. Numerous calcium determinations were done with the calcium remaining elevated and as high as 14.8 mg%. Parathormone level was 3 nanograms (normal values 0 to 1.5 nanograms per ml.)

The patient was given Mithramycin for his hypercalcemia and had a neck exploration. A parathyroid adenoma was found, consisting of a well demarcated nodule of soft tan tissue measuring 2 cm. in largest dimension, with a rim of normal parathyroid tissue. Biopsies of the other parathyroid glands were normal. The patient later underwent a 75% partial gastrectomy and your slides are from that specimen.

The specimen consisted of a 7 x 12 cm. piece of stomach which was covered by multiple brown tan polyps. Also submitted were four polyps removed from the unresected fundus of the stomach and these were benign.

The followup on this patient is that his serum calcium is normal 8.9, and that his weight has gone from 135 to 154 pounds. The patient is receiving vitamin B12 shots monthly and has no symptoms of diarrhea or "dumping".

Submitted by: Pathology Staff
St. Paul Ramsey Hospital

CASE #5

This 42 year old white female presented to another hospital with abdominal pain, vomiting and constipation. Exploratory laparotomy revealed marked peritonitis and areas of gangrenous small bowel. The patient had a primary ileostomy but later went into renal failure. She also had a tracheostomy for respiratory failure. The patient was transferred to Ramsey County Hospital and Medical Center. A second exploratory laparotomy revealed more gangrenous small bowel which was removed. The patient then went downhill with her course characterized by episodes of sepsis, hypotension, electrolyte imbalance, anemia, neutropenia, thrombocytopenia, hypocalcemia, and DIC. Blood cultures were persistently negative. Peritoneal cultures revealed Enterococcus. Sputum cultures revealed yeast and gram negative rods. The slides submitted are from the segment of small bowel removed at St. Paul Ramsey Hospital (second operation). The specimen measured 85 cm. in total length, and was markedly discolored, especially on the antimesenteric surface, and had patchy areas of greenish-black fibrinous exudate on the serosal surface.

Submitted by: Pathology Staff
St. Paul Ramsey Hospital

CASE #6

This 16 year old male was admitted to St. Paul Ramsey Hospital with a one month history of a 15 to 20 pound weight loss and crampy, intermittent epigastric pain unrelieved by antacids. During the week prior to admission he had been vomiting and anorectic. On admission the physical examination was unremarkable. Laboratory examination revealed: hemoglobin 13.4 grams with mild microcytosis, WBC 10,100 with 66% PMN's, uric acid 9.4 mg%, total protein 8.7 gms%, with elevation of all globulins by ELP. Stool guaiac was 4+. Barium enema revealed an ileocolic intussusception, and a laparotomy was performed. A 7 cm. soft, fleshy tan tumor was found in the distal ileum.

Submitted by: Pathology Staff
St. Paul Ramsey Hospital

CASE #7

This 68 year old man presented with a vague nagging feeling in his right flank for approximately 6 weeks. He noted that his appetite had decreased but there was no definite weight loss. He denied the presence of bloody or tarry stools; however, the guaiac was 3+ at the time of his first office visit. He was admitted to the hospital and the workup revealed a microcytic hypochromic anemia, and an "intrinsic mass" at the ilio-cecal valve. A laparotomy was performed and the distal ileum plus the entire cecum, appendix, and ascending colon to the hepatic flexure were removed. Three separate lesions were identified at gross examination. The most remarkable was a raised fungating firm mass at the ilio-cecal junction. On microscopic examination this was seen to be a typical infiltrating adenocarcinoma and this lesion is not included in the seminar slides. There was a second polypoid mass 2 cm. distal to the ilio-cecal valve within the ascending colon. This measured 2 x 2.4 x 1 cm. and was attached by a broad based stalk. This lesion is included in the seminar slides. The third polypoid lesion was noted at the hepatic flexure, 3 cm. from the distal line of resection. The mass measures 2 x 2 x 1.2 cm. and had a 1 cm. broad stalk. This lesion is included in the seminar as the slide labeled "B".

Submitted by: Central Regional Pathology Laboratory
St. Paul, Minnesota

CASE #8

This 37 year old male was admitted with a chief complaint of midabdominal pain associated with nausea. There was point tenderness in the right lower quadrant and an appendectomy was performed. At gross examination, the appendix was 7 cm. in length and had a variable diameter, from 4 mm. to 1.5 cm. At the distal tip there was a bulbous enlargement and sectioning through that area showed the wall to measure up to 5 mm. in thickness. The wall within this area was firm and white and the serosal surface was glistening and smooth.

Submitted by: Central Regional Pathology Laboratory
St. Paul, Minnesota

CASE #9

This 23 year old man had a 15 year history of "probable" ulcerative colitis. He had been receiving therapy with Asulfadine. In 1970 the patient was deferred from military service because of his gastrointestinal problem. Three months prior to the present admission he began to have episodic crampy abdominal pain and diarrhea. Barium enema showed "involvement" of the entire colon with ulcerative colitis. The patient then began having increasing right lower quadrant pain associated with nausea and vomiting. A barium enema showed total obstruction of the transverse colon. The patient underwent a subtotal colectomy with permanent ileostomy.

Submitted by: John Stoltenberg, M.D.
Hennepin County General Hospital
Minneapolis, Minnesota

CASE #10

This 25 year old lady had an "anal fissure" removed but experienced persistent difficulty. The surgical excision site would not heal and several months later she was readmitted for re-excision of this fissure. Past medical history revealed no serious illnesses or surgery. However, there was a vague history of "increased frequency of bowel movements". 2 slides.

Submitted by: Aina Galejs, M.D.
Midway Hospital
St. Paul, Minnesota

CASE #11

This unfortunate but amazing 49 year old man has been a resident of the operating room at St. Paul Ramsey Hospital for the past two years. He was born with a meningomyelocele which was closed during the first year of his life. However, he had marked muscular weakness in his legs plus a paralyzed bladder for all of his life. At the time of late childhood (perhaps) or during his midteens (for certain) he has had a slowly enlarging "papilloma" of the left buttock and natal cleft. This slowly enlarged and became a draining fungating problem until 1968 when it was excised in a somewhat piecemeal fashion. The diagnosis at that time was "invasive squamous cell carcinoma" but there was apparently some disagreement about this diagnosis. The lesion was again resected in 1972 and this was followed by skin grafting. However, the tumor again recurred and in July, 1973, he was admitted to St. Paul Ramsey Hospital. At this time he had a huge fungating mass of the buttocks, perianal skin, and anus. At this time he had an exenteration of the rectum, bladder and prostate and a "new" diagnosis was offered. Since that time the patient has had two additional local surgical procedures for "recurrence", one in February and one in March of 1974. There has also been a chronic problem with wound healing. Nonetheless, the patient has managed to spend a small amount of time away from the operating theater at St. Paul Ramsey Hospital and during these intermissions he has managed to maintain his employment, as he has done for his entire adult life.

Submitted by: Pathology Staff
St. Paul Ramsey Hospital

CASE #12

This 80 year old man presented with a "tumor" of the anal skin. Upon further examination by the local surgeon the tumor apparently arose from skin but it grossly extended into the anal canal. There was superficial ulceration and the pre-operative diagnosis was "transitional cell carcinoma".

Submitted by: Ralph Tarnasky, M.D.
St. Alexius Hospital
Bismarck, North Dakota

CASE #13

This 15 year old girl was seen in the emergency room at St. Paul Ramsey Hospital for "difficulty swallowing". She complained of midsternal pain and left upper quadrant pain which was exacerbated by eating and relieved by sitting up. The patient has had an "enlarged spleen" since she was 6 weeks old; the patient has sickle cell disease (SA) but has not suffered from anemia. Her hemoglobin at the time of admission was 12 grams with a normal white count and differential. The patient was admitted for investigation of the substernal pain and dysphagia. A \$990 radiographic workup followed with the conclusions of "possibility of left renal tumor, large extrinsic gastric mass probably enlarged spleen, negative chest film, enlarged spleen by ultrasound with the remote possibility of a mesenteric cyst, spleen size within upper limits of normal by splenic scan, and a filling defect of the liver by liver scan". Following these clear cut impressions from Radiology, the patient had a laparotomy. At the time of this procedure two separate masses were noted. There was a 10 x 8 x 8 cm. mass within the wall of the greater curvature of the stomach. This was removed with a portion of the surrounding stomach wall including the gastric mucosa. The mass was generally rounded and consisted of a cyst which contained clear watery fluid. Most of the wall appeared to consist of muscle with a mantle of thick glassy hyalinized material on the peritoneal surface. Upon microscopic examination, this cyst was seen to be lined by a single layer of columnar and pseudostratified columnar nonciliated epithelium. This epithelium was compatible with "enteric" epithelium and the lesion was diagnosed as a gastric duplication. The second nodule was removed from the upper surface of the left lobe of the liver, between the diaphragm and the liver. This consisted of a 15 gram rounded nodule of tissue measuring 3 x 2 x 2 cm. The tissue was glistening, firm and gray-white with several small cystic areas noted. These cysts contained clear fluid. Sections for the seminar case are taken from this subdiaphragmatic or suprahepatic nodule.

Submitted by: Pathology Staff
St. Paul Ramsey Hospital

- CASE 1. My Dx: Benign lymphoepithelial lesion of salivary gland. Templeton was worried about malignant lymphoma because of the unilaterality, but ended up calling it benign.
- CASE 2. My Dx: Leiomyosarcoma of stomach. Templeton agreed. The people at St. Paul-Ramsey were trying to make a hemangiopericytoma out of it.
- CASE 3. My Dx: Leiomyoma of stomach. Templeton agreed, although expressed he had a great deal of difficulty to tell it apart from neurofibroma. The patient comes from a Neuropsychiatry institute, but does not have Von Recklinghausen's disease.
- CASE 4. My Dx: Hyperplastic polyp of stomach (in a patient with MEA). Templeton agreed. Pointed out the presence of eosinophiles, but he did not know what to do with them.
- CASE 5. My Dx: Necrotizing enteritis with fungi present. Looks like Mucor but there are septae. Templeton called it Mucormycosis. The surgical diagnosis was hemorrhagic infarct of bowel due to postoperative fibrous adhesions with superimposed fungal overgrowth. The patient died but there was no autopsy. The organism was not cultured.
- CASE 6. My Dx: Malignant lymphoma, PDL with plasmocytoid differentiation. Templeton called it PDL. The patient came back with a tumor in the anterior abdominal wall that has a similar microscopic appearance. Templeton mentioned the possibility of American Burkitt's lymphoma but he discarded it. He thinks that so-called American Burkitt's is not Burkitt's.
- CASE 7. My Dx: Two adenocarcinomas, one arising in a villoglandular polyp. Templeton agreed. The discussion centered on the malignant potential of polyps. Templeton mentioned that Gilbertsen has reduced the risk of polyp patients to develop colon cancer 90% by taking out the polyps.
- CASE 8. My Dx: Mucinous carcinoid. Templeton agreed.
- CASE 9. My Dx: Well-differentiated adenocarcinoma (in a patient with ulcerative colitis). Templeton called it colitis cystica profunda. John Coe mentioned that the glands extended into the mesentery, but there were no lymph node metastases. The patient is ok 4 months following surgery.
- CASE 10. My Dx: Non-caseating granulomas of anus, R/o Crohn's disease. Templeton agreed. The contributor mentioned that this patient indeed has Crohn's disease of the terminal ileum.
- CASE 11. Well-differentiated squamous cell carcinoma of skin, related to verrucous type. Templeton agreed with squamous cell cancer. The DX at St. Paul-Ramsey is verrucous carcinoma.
- CASE 12. Templeton called this true basal cell carcinoma of the perianal skin. The obvious differential diagnosis is basaloid (cloacogenic) carcinoma. I think he is probably right.
- CASE 13. Pulmonary sequestration between diaphragm and liver. Templeton agreed.

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MIDWINTER ANATOMIC PATHOLOGY SEMINAR

Diseases of the Gastrointestinal Tract

Dr. Templeton's Diagnoses

1. Benign lymphoepithelial lesion, parotid gland.
2. Leiomyosarcoma, stomach.
3. Degenerating leiomyoma, stomach.
4. Hyperplastic polyps, multiple, stomach.
5. Mucormycosis, bowel.
6. Malignant lymphoma, poorly differentiated, diffuse, small bowel.
7. Polypoid adenocarcinoma, two separate lesions, with one arising in a mixed polyp.
8. Carcinoid tumor, mucinous type, appendix.
9. Colitis cystica profunda occurring in ulcerative colitis.
10. Regional enteritis presenting as anal fissure.
11. Squamous cell carcinoma, low grade, anus.
12. Basal cell carcinoma, arising in perianal skin.
13. Pulmonary sequestration occurring in association with a gastric duplication.

DISCUSSION OF CASES

1. The differential diagnosis in this case centered around the question of whether or not this is a benign lymphoepithelial lesion or whether this was a malignant lymphoma presenting as a salivary mass. The majority of the participants felt that this was a benign lymphoepithelial lesion and there were questions concerning the possibility that this patient had Sjogren's syndrome or "Mikulicz's Disease". This patient had been re-examined intentionally with those thoughts in mind and there was no clinical evidence to support those considerations. The other parotid gland was not clinically enlarged but Dr. Rosai suggested that even though the opposite gland was not enlarged there still may have been a lymphocytic infiltrate microscopically within that gland. The patient is doing well.
2. The majority of participants felt that this lesion was a malignant smooth muscle tumor and Dr. Templeton was kind enough to agree with the consensus opinion. However, several people suggested the diagnosis of leiomyoblastoma and this was considered by the group in their discussion. I believe that this diagnosis was ruled out on the basis of the multiple blocks which we examined within which we found obvious muscular invasion and with the showing of those photographs at the conference the people who were considering leiomyoblastoma as a diagnosis were quietly ushered out of the room. Dr. Templeton was concerned "why" a 17 year old patient should develop a leiomyosarcoma of the stomach and brought up the broad subject of immunology and immunologic defenses against malignancy. He had noted lymphocytes around many of the tumor lobules. The patient is doing well at this point.
3. The diagnoses in this case were split rather evenly between neurofibroma and leiomyoma. Dr. Templeton nicely straddled the fence and said that it could be either but since it was occurring within the stomach he would favor the diagnosis of leiomyoma simply because leiomyomas were more frequent than neurofibromas within the stomach. He became exceedingly nervous when it was revealed to him that the pathologist who submitted the case has a strong involvement in neuropathology and Dr. Templeton was paranoid enough to consider the possibility that this patient might have von Recklinghausen's Disease and that the diagnosis would then be a neurofibroma. We were not given the information that this patient did have neurofibromatosis but the diagnosis of the submitting pathologist (Dr. Obert) was neurofibroma and there is no doubt that a case can be made for that.
4. Many of the participants in the conference felt that this lesion represented a true adenoma or adenomatous polyp of the stomach. Dr. Templeton felt that this was a regenerative of hyperplastic polyp and the staff at St. Paul-Ramsey Hospital who submitted this case also felt that these were not true adenomatous polyps but were hyperplastic or regenerative nodules. The lengthy history of hyperparathyroidism with a parathyroid adenoma was given to you in a direct attempt to mislead; there is no published report that we know of concerning parathyroid adenomas and multiple hyperplastic benign regenerative polyps of the stomach. This entity shall be known as the Templeton syndrome.
5. This case of mucormycosis of the bowel was diagnosed by everyone. It was included in the seminar because we wanted to broach the general subject of fungal diseases of the gastrointestinal tract and bring up the question of secondary fungal infection in debilitated patients. This patient expired before any type of immunologic investigation could be undertaken and we were unable to obtain an autopsy. However, Dr. Templeton filled us in on his wide experience in infectious diseases in Africa and also mentioned the possibility of primary mucormycosis of the intestinal tract occurring in previously healthy patients.

6. Everyone diagnosed this as a malignant lymphoma. The general subject of lymphomas was discussed. This patient is doing poorly; two weeks ago he presented with subcutaneous lymphomatous nodules of the abdomen.
7. This case was presented to initiate discussion of the general subject of malignancy occurring in polyps of the colon. Proper sectioning of the blocks were extremely difficult in this case but with material at our disposal we were able to determine that this patient had a run of the mill infiltrating carcinoma occurring at the ileocecal valve, a polypoid adenocarcinoma at the hepatic flexor, and a carcinoma arising within a mixed adenoma in the ascending colon. In addition, the patient had a fourth lesion within the transverse colon and this was not removed in surgery. We had all seen better examples of tumor arising within mixed villous and adenomatous polyps but it is difficult to make large numbers of slides all showing the same thing.
8. This case was diagnosed by many of the participants as an adenocarcinoma of the appendix. It may well be an adenocarcinoma of the appendix but the histology matches the lesion described as a "Goblet cell" or mucinous carcinoid tumor. This entity is described in Cancer, March, 1974, and Cancer, August, 1974. The tumor has features both of a carcinoid and a carcinoma and apparently there is some question about the exact nature of this tumor.
9. This lesion incited the most discussion at the seminar. Obviously, a large number of people felt that this was adenocarcinoma complicating ulcerative colitis. Dr. Coe and the group at Hennepin County Hospital had diagnosed this as an adenocarcinoma but had "second thoughts" about it. They had this as a problem on frozen section and although it was called adenocarcinoma, they strongly considered the possibility of colitis cystica profunda. Since Dr. Templeton did not have to sign the surgical report, he, in his inimitable fashion, called it benign (colitis cystica). He was in good company since many of the residents present also felt that it was benign. There is a very short follow-up on this patient and there is no evidence of disseminated disease.
10. The differential diagnosis in this case was Crohn's Disease versus granulomatous proctitis versus other granulomas such as TB and fungal infections. The submitting pathologist, along with Dr. Templeton and many other pathologists, felt that this was a harbinger for regional enteritis and it turned out that this patient did in fact have regional enteritis.
11. The question on this case was whether or not the patient truly had squamous cell carcinoma and if the patient did have carcinoma, what was the background for this tumor. Dr. Templeton showed slides and discussed several cases of squamous cell carcinoma and other epithelial lesions of the skin arising in the area of chronic inflammation, particularly tropical ulcers, which he had seen in Africa. We felt that this was squamous cell carcinoma, but in particular, we felt that this was arising as a verrucous carcinoma or a giant condyloma. The giant condyloma does occur in the genital, perineal and anal region and current thinking on the giant condyloma is that it is a verrucous squamous cell carcinoma. We have put this lesion into that category and follow up seems to support that diagnosis. The patient has had several local recurrences but no evidence of metastatic disease.

12. There was divergence of opinion on this case with many participants diagnosing this lesion as a transitional cell or cloacogenic carcinoma. Dr. Tarnasky had originally made that diagnosis but subsequently had amended the diagnosis to basal cell carcinoma of the anus. This is not to be confused with a "basaloid" carcinoma which is a transitional cell carcinoma. Dr. Templeton was curious as to the etiology of basal cell carcinomas of anal skin. Most basal cell carcinomas of the skin arise on sun exposed areas and the anus, under usual circumstances, is not a part of the body usually exposed to sunlight. However, Dr. Templeton solved the entire problem by noting that in England there are certain snooty or stuck-up people who believe that the sun shines from their arses. This would nicely explain the occurrence of basal cell carcinoma in that area.

13. This case was submitted as an overt curiosity. The patient had the laparotomy because of the gastric mass which turned out to be a gastric duplication. The filling defect of the liver turned out to be the wad of cartilage and intact lung, leading to the diagnosis of pulmonary sequestration. Pulmonary sequestration is not an uncommon lesion in infants but it usually occurs within the thoracic cavity. Its occurrence as an extra pulmonary lesion is distinctly rare and its association with gastric duplication is nearly unique. A few hemosiderin laden macrophages were present in the alveolar spaces and Dr. Templeton wondered if the patient was in heart failure as well. Dr. Ward from Mount Sinai brought along a recent case of a pedunculated extramural gastric duplication and mentioned that at least this case was one that he got right but he had been applying negative oscultory pressure to a posterior mammary gland on several of the earlier seminar cases.