

Lecture N. 24

Diseases of the Ileocecal Appendix

In the preceding Lecture (No. 23) the appendix is the protagonist of numerous surgical situations that are often difficult to diagnose and treat, even if the role it plays is secondary to development disorders of the colon. As such, the organ may be found in any one of a number of positions in the abdominal cavity depending on the location of the cecum.

Under normal conditions, the appendix lies in the right iliac fossa, planted at the point where the three taenia coli meet on the medial face of the cecum, 2 - 3 cm. below the ileocecal valve. But it may also extend in any of a number of ways: descending, reaching as far as the pelvis; ascending, clinging to the posterior wall of the cecum (retrocecal appendix - Fig. 1); medial, towards the abdominal cavity; lateral, between the cecum and the lateral abdominal wall. These varying positions that under normal conditions the appendix may assume are able to influence the anatomic and clinical pictures of the appendicular disease.

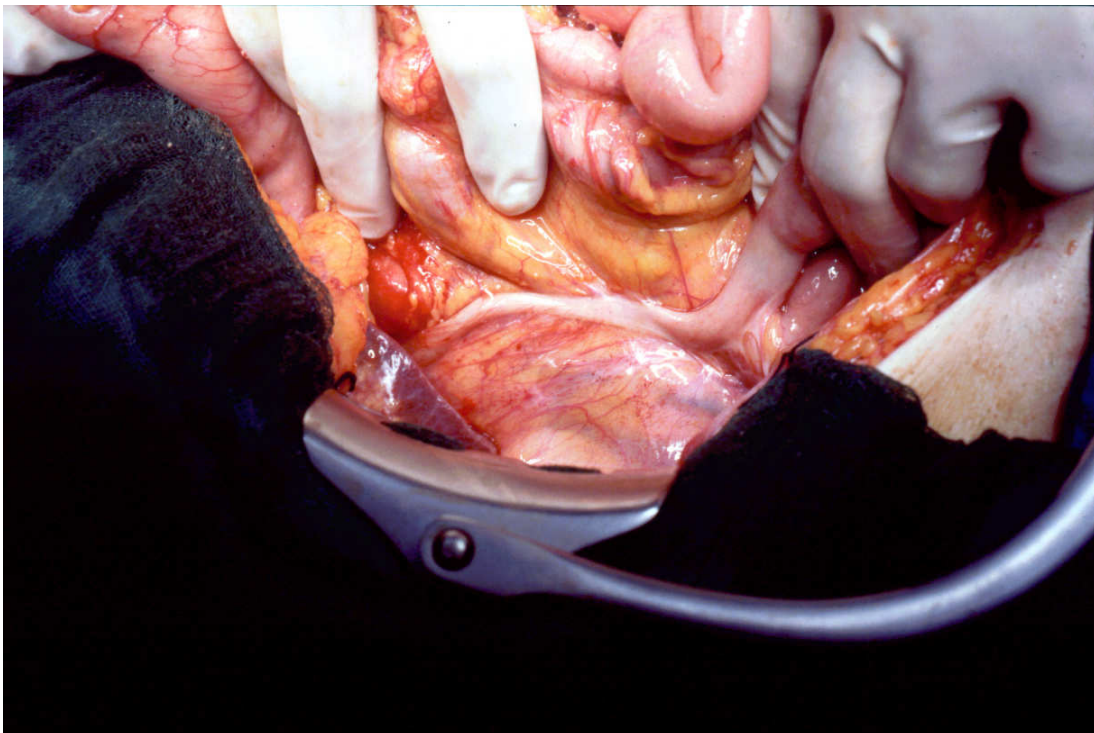


Fig. 1

Long retrocecal appendix reaching the subhepatic region

The structure of the appendix mimics that of the colon: serosa, muscular, Auerbach's and Meissner's myoenteric plexi, muscularis mucosae and mucosa. It differs for the large amount lymphatic tissue, which actually constitutes areas of true and proper lymphatic follicles. These are often separated from the appendicular lumen by a single layer of epithelial cells, which sink to form tubular crypts. Such an arrangement resembles that of the palatine tonsil, and, indeed, this similarity has prompted the appellation of "*abdominal tonsil*" This abundance of lymphatic tissue is characteristic of infancy and adolescence.

The vascular and nervous structures serve the appendix through the mesoappendix: the appendicular artery, a branch of the superior mesenteric-ileocolic artery, is terminal; the veins are tributaries of the ileocolic-portal system vein; the lymphatic vessels drain the pericecal lymph nodes, and from here, via the superior perimesenteric vessels, lymph reaches the peripancreatic lymphatic structures. These vascular details, as we will see below, corroborate the possible long-term complications of appendicular diseases.

Nerve fibers connected to the celiac plexus through the superior mesenteric plexus also pass through the mesoappendix. This particular anatomic feature will arise again in the section devoted to the pain symptomatology of the appendix.

Appendicitis

Two clinical pictures are described for the disease: acute and chronic.

Acute appendicitis is the most frequent cause for emergency abdominal surgery. The percent incidence of the disease in the general population has been estimated to lie between 1/500-600 inhabitants. The condition strikes above all during infancy and adolescence, but also occurs in all other ages.

From a pathogenetic standpoint, the infection is nearly always endogenous and is sustained by common habitual guests of the intestine, such as coli, staphylo-, streptococcal, and much less often, anaerobic bacteria. This bacterial flora can easily accumulate in the above-mentioned tubular crypts of appendiceal mucosa, thereby creating - if virulent - a unhindered reaction by the adjacent lymphatic component and inflammatory compromission of the entire wall. Such reactivity will, of course, be more intense where this component is more greatly represented, i.e., in younger subjects. It seems obvious that, if the appendiceal lumen becomes obstructed, the closed chamber that results will fatally promote bacterial multiplication and induce endoluminal hypertension, stasis and ischemia with consequent parietal invasion of germs. Obstruction may arise for a number of reasons: angulation or torsion of the organ; a foreign object (fecaliths, a cherry pit, parasites); hyperplasia of lymphatic tissue; sclerotic retraction from previous inflammatory events.

Under particular circumstances, the “abdominal tonsil” may, especially in children, recall via a hematogenous route circulating germs from distant inflammatory sources.

The pathologic anatomy distinguishes four forms of acute appendicitis - catarrhal, purulent, phlegmonous and gangrenous - which usually represent increasingly worsening evolutionary stages of the disease. Each of these forms may, however, present alone from disease onset.

- Catarrhal appendicitis - hyperemia, edema, lymphatic hyperplasia, leukocytic infiltration of the mucosa and submucosa with endoluminal sero-leukocytic exudate. If appendiceal canalization is impeded, the exudate expands the organ, thereby constituting a **hydrops of the appendix**.
- Purulent appendicitis (Fig. 2) - the organ is increased in volume, rigid and inflamed due to congestion. Inflammation extends to the lamina muscularis mucosae and the serosa, and the mucosa, thickened and congested, often presents ulcerations that can reach the serosa and create perforations. Obstructed canalization of the organ leads to **appendiceal empyema**.
- Phlegmonous appendicitis, in keeping with the definition of phlegmon, presents extended compromission of all layers of the organ, with numerous abscesses in the wall and abundant fibrinopurulent exudate on the serosa. Inflammation also involves adjacent structures: the cecum, the last ileal loop and the contiguous parietal peritoneum. As such, it constitutes a fibrinopurulent peritonitis limited to the right iliac fossa. This circumscribed peritonitis represents the anatomic basis of the so-called **ileocecal plate**.
- Gangrenous appendicitis - since, as we've said, the appendicular artery is a terminal structure, any thrombotic event will naturally lead to total or partial necrosis of the organ. Thrombosis may arise in the most serious forms of obstructive appendicitis, and is promoted by the necrotizing action of anaerobic germs or also by the presence of an endoluminal foreign object. The event evolves into the breakdown of the wall, perforation, consequently, peritonitis.

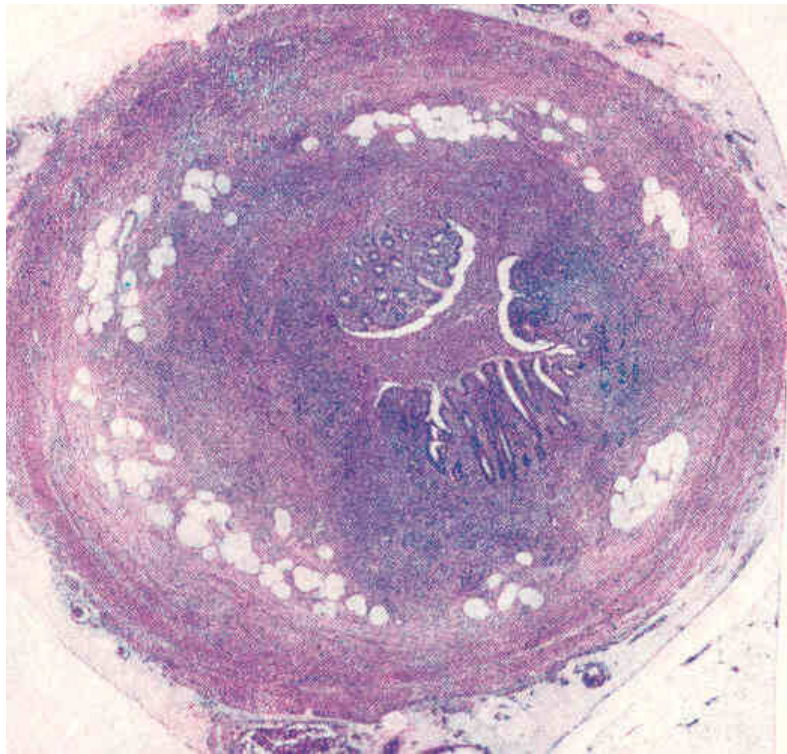


Fig. 2

Acute purulent appendicitis - pus in the lumen, inflammatory infiltration of the wall

The acute state of appendicular inflammation inevitably impacts on the peritoneum. This may range from a modest state of “peritonism” (a term corresponding to the semiotic sign of Blumberg), that is the simple sharing of the peritoneal serosa, to outright peritonitis, which may be widespread or localized. Circumscribed purulent peritonitis is defined as an **appendiceal abscess**. As the term implies, it is situated in the vicinity of the appendix, from which it draws its name. In most cases, the abscess will be located in the right iliac fossa, but if the appendix is in an anomalous position, it could form elsewhere: the pouch of Douglas, the pelvic excavation, the subhepatic region, the left iliac fossa, etc.

An ileocecal abscess may affect the retrocecal, and hence, the retroperitoneal, spaces, thereby giving rise to a **retroperitoneal adenophlegmon** with possible involvement of the psoas muscle (psoitis). Micotic emboli resulting from the acute inflammation of the appendix may reach the liver via the portal vein, thus inducing suppurative phenomena, such as, for instance, a hepatic abscess.

The clinical picture of acute appendicitis is fundamentally that of acute abdomen, a syndrome that requires careful evaluation in view of a more than likely surgical urgency.

The symptomatology of acute appendicitis is generally characteristic and constitutes the onset and often the key to interpret the diagnostic work-up. This begins with gathering subjective symptoms, which normally are:

pain, nausea, vomiting, closed bowels, fever.

- Abdominal pain - generally arises in the right iliac region and is violent, at times with an onset that resembles paroxysmic colic and may be preceded by prodromic disturbances, such as dyspepsia, nausea and bowel irregularity. Pain may irradiate to the thigh or to the lumbar or gluteal region. It may begin in the epigastric or periumbilical region, and in these cases, induce some diagnostic errors.
- Nausea and vomiting - vomiting may at outset be alimentary, may become biliary, and in the case of peritonitis, may become fecaloid.
- Closure of the bowels - to gas and feces.

- Fever and tachycardia - fever is usually high (38-39°). Tachycardia may arise earlier, at times disproportionate to axillary temperature, which instead is normal or slightly high; in these cases, however, rectal temperature will be high, even markedly so, and will correspond to pulse activity.

Physical examination is the next step and usually provides the key to interpreting the disease.

- Inspection - in forms with peritoneal compromise a reduced respiratory expansion of the abdomen may be seen. When invited to cough, the patient complains of accentuation of right iliac pain that induces him/her to place a hand where pain is greater. More serious forms will compel immobile posture with pain-relieving flexing of the lower limbs.
- Palpation - the greater the severity of the disease process is the more a defensive muscular contraction will become. This may be limited to the lower right quadrant if the appendicular and peritoneal inflammation is modest, or spread to the entire abdominal wall if peritoneal compromise is more serious. Lest intense pain be caused, palpation must be superficial. At times even lightly brushing the skin causes pain (cutaneous hyperesthesia).

Peritoneal compromise in the initial stages of simple peritonism is detectable with Blumberg's maneuver (Blumberg's sign). Only in milder forms of catarrhal appendicitis can palpation reveal the iliac localization of pain.

- Auscultation - peristaltic silence.
- Rectal exploration - vaginal if possible and indicated -

Extreme tenderness to the right

Possible inflammatory involvement of the pelvic cavity or the pouch of Douglas.

Since it is an acute inflammatory process, laboratory tests will define the general reaction to the event. The greater the inflammatory aggression is the higher the degree of neutrophil leukocytosis will reach. And likewise all of the other well-known signals: ESR, CRP, etc.

Time ago, an option to treat cases with a mild symptomatology, usually corresponding to a catarrhal appendicitis, was "cooling", which entailed bed rest, ice packs, special diet and, above all, careful observation so as to avoid worsening. In fact, in attenuated cases symptoms may resolve spontaneously: the bowels open again to gas and feces, nausea and vomiting disappear, as do fever, pain, muscular defense and leukocytosis. Today this process may be expedited with antibiotics. This neglects the fact, however, that modern minimally invasive surgical techniques can resolve the problem relatively easily and quickly. Moreover, even after a possible spontaneous resolution, the appendix will sooner or later make itself felt again and will require a surgical solution.

More serious forms of acute appendicular disease necessarily compel emergency intervention, which is generally preceded or accompanied by antibiotic therapy. In the past above all and at times today, a delay in providing surgical care may give rise to the formation of the *ileocecal plate*, appendicular abscess or generalized peritonitis.

In Lecture No. 23, we spoke at length about the possible anomalies that may affect the organ in question. As such, the symptomatology described above may change, thereby implicating the need for differential diagnosis. But the question is more theoretical than practical, because it is the clinical picture, that is acute abdomen, that draws the attention of the examiner. We are dealing, in fact, with acute appendicitis, be it in its varying degrees of severity. It will therefore be this state of the abdominal disease that will dictate management. And for more than 20 years, and even more so today, this entails the video-laparoscopic approach, which enables the definition of the diagnosis through direct exploration of the abdominal cavity, the subsequent localization of the source of inflammation - wherever it may be - and the related therapeutic provisions to implement.

Therapy of acute appendicitis is surgical and entails appendectomy, which is usually performed under emergency circumstances. All the more so for the most serious forms and those affecting the peritoneum. In these instances, thorough toilette of the peritoneum and treatment of possible abscessual accumulations, which necessarily require external drainage, obviously become indispensable. Laparotomy, a legacy of traditional surgery, takes advantage of a small access in the right iliac region via a number of techniques (according to Stropeni, McBurney, etc.) in uncomplicated cases. On the contrary, the more serious involvement of the peritoneum is, the larger the access will become. When the source of inflammation is in an entirely different site, as in cases of an anomalously located appendix, laparotomy will be performed accordingly (subhepatic, median, left quadrants). As we have said and as is widely known, video-laparoscopy has allowed surgeons to overcome most of the difficulties - many of which were extremely serious - that for so many years handicapped their treatment of this condition, and likewise and above all, to relieve patients of surgery-related traumas.

Chronic appendicitis

There are two types of disease, primary and secondary. Recognizing the latter and implementing its treatment are not difficult: it is, in fact, secondary to a previous acute appendicular event, which clearly emerges from the patient's medical history, that went untreated or undertreated and hence progressed into a chronic inflammation.

Primary chronic appendicitis, on the other hand, is a pathological condition with its own anatomical features. From an etiological standpoint, much debate has centered on the true primary nature of the process, with some authors arguing that - at the outset - there is an acute event with little symptomatologic meaning that went undiagnosed as such, resolved spontaneously and, as a result, was never even subject to treatment. Nevertheless, the disease is relatively frequent and also has its own unique anatomic-clinical attributes. Macroscopically, evident and varying alterations can be found. The organ may be rigid, sclerotic, at times threadlike, adhering to nearby structures (cecum, mesentery, greater omentum, female organs) that may be involved in the inflammatory process, above all the last ileal loop and the cecum (perityphlitis). Or it may appear swollen with a thickened mesoappendix and satellite lymphadenopathy. Obliteration of the appendicular lumen is also somewhat frequent, at times with the presence of foreign bodies (fecaliths, parasites, grape seeds, etc.). The histology corresponds to these pictures: chronic inflammation characterized by extensive leukocyte infiltrates, serous exudation, lymphatic hyperplasia (which may at times be multinodular), signs of sclerosis, and so on.

The symptomatology is fundamentally characterized by pain, which may be continuous, or occasional with varying intermittence. Classically, pain is located in the right iliac region and may irradiate to the ipsilateral thigh or, more rarely, to the lumbar region, which clearly makes differential diagnosis more problematic. Quite often, various other disturbances are associated with pain: dyspepsia, hyporexia, unequivocal anorexia, nausea, stypsis and diarrhea (often alternating), mild fever. This possible variety of symptoms can raise diagnostic issues and doubts.

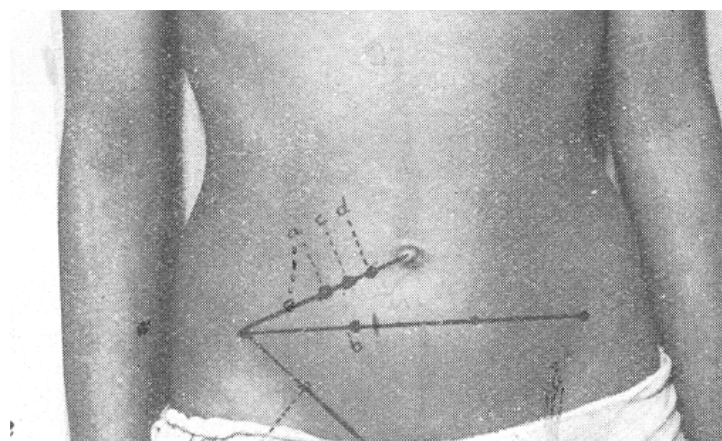


Fig. 3 - a) McBurney, b) Lanz, c) Morris, d) Munro, e) Jalaguier

Chronic appendicitis, in contrast to acute disease, allows the practitioner to apply the classic rules of semiotics. On palpation, the following will become pathognomonic: McBurney's, Lanz's, Morris', Munro's and Jalaguier's points of tenderness (Fig. 3); and Aaron's, Rowsing's and Labinskji's signs/diagnostic maneuvers (see also in this site: Dispense di Semeiotica Chirurgica e/o Synopsis di Patologia e Clinica Chirurgica).

Here, perhaps more so than in acute syndromes, an anomalous position of the appendix will significantly impact on the location of pain (which nonetheless is still accompanied by the array of symptoms described above). If the organ is subhepatic, or worse still located in the left quadrants of the abdomen, diagnosis will be impossible without instrumental support. Not infrequently, patients affected by chronic appendicitis complain of epigastralgia. This is often seen in young school-aged patients. The following scenario is familiar: the child is accompanied to school, where, especially in the first few days, he/she complains of epigastralgic pain ("I have a tummy ache"); the teacher becomes worried, the mother is notified, the child is taken home, where the symptoms gradually disappear. The first hypothesis is that this is a reflex stemming from nervous tension or apprehension that the child experiences when faced with a new or feared event. And this may well be the case. But recurrent epigastralgia when these conditions do not exist justifies the suspicion of a possible appendicopathy. All the more so seeing that the disease occurs at a very high rate in children.

In women, gynecological disturbances may coexist: clearly these demand inspection to verify this eventual relationship in contrast with the presence of two possible concomitant problems.

Appendicular disease represents a source of neurovegetative stimulation that varyingly affects the entire abdomen, in particular the colon, above all if protracted in time. The most common consequence is colic dyskinesia. The bowels become irregular, but tend to become stytic; pain affecting all segments of the colon may arise; ultimately, a picture of so-called irritable bowel or even "spastic colitis" becomes evident. Appendectomy generally resolves the problem, which can, however, remain if the situation persists over time.

The diagnosis of chronic appendicitis requires instrumental means when, as mentioned above, the clinical picture does not reliably allow it based on semiologic findings alone. Here ultrasonography, which can rule out suspected gallbladder, pyeloureteral or adnexal disorders, may come into play. Radiography, too, via a barium swallow, can confirm diagnosis: missing or partial opaquing of the appendix, onset of localized pain on compression of the appendix, and an opaque enema are helpful in cases of suspected anomalies of position.

Therapy of chronic appendicitis is surgical. As for acute appendicitis, appendectomy is the treatment of choice for the chronic form, as well, but, unlike the latter, it is elective. This also holds, in our view all the more so, for primary forms. As said earlier, chronic appendicular disease impacts negatively on digestive system function, particularly on the colon. It follows that localized disease may spread, leading to more serious outcomes. Appendectomy, therefore, should be performed promptly, before this picture develops. Delayed intervention may not always remedy damage already done. This is why resorting to conservative therapies, generally medical-dietary (antispastic agents, mild laxatives, diets, etc.) or another type, as some medical circles suggest, is not advisable.

From a technical standpoint an appendectomy can be performed with a laparotomic approach through limited incisions, as with the McBurney or the McBurney-Stropeni procedures. Above all, this latter (which we prefer) implements an incision that is more horizontal, very limited and slightly more advantageous aesthetically. With this technique access to the peritoneum entails only a contained opening of the aponeuroses of the large oblique muscle and the simple dissociation of fibers of the small oblique and transverse muscles. This approach is indicated if the surgeon expects to find an appendix in its normal position and without excessive pathologic alterations.

If, on the other hand, there is a presumed need for wider access, a more vertical incision that can more easily be extended, such as a McBurney, should be used. Otherwise, resorting to a different technique is advisable: a Jalagueir or a Roux pararectal laparotomy (low, oblique and transversal - if exploration of adnexa of the uterus is indicated).

Surgical textbooks suggest many different approaches to the ileo-cecum-appendicular organ complex, depending on the conditions encountered: these details lay beyond the scope of this lecture. One particular, however, should be highlighted: once the peritoneum has been opened and the anatomopathological conditions of the appendix have been assessed, the last 20 centimeters of the terminal ileum should be extracted, because, and above all if the appendix seemingly shows none of the alterations that were expected, the symptomatology could have been caused by the inflammation of a Meckle's diverticulum (meckelitis), which, in such a case, should be removed.

Careful examination of the features of the excised organ is always very important, since (as we will see below) it may be affected by various lesions. One of these that could have negative secondary effects if not recognized is Crohn's disease. In the first stages this disorder does not present the well-known morphological characteristics; indeed, it may reveal itself by giving affected organs only a pale red color, a feature that may go unnoticed by the surgeon. In these cases, quick onset suppuration of the wound is common. For these reasons histopathologic examination of the removed appendix should always be performed.

A personal clinical case. One day - one night rather - I received an emergency call for a strangulated right inguinal hernia. All the symptoms of an acute condition were present: irreducible hernia, pain, peritonism, closed bowels, sepsis. Surgery was performed immediately. The hernial sac appeared extremely inflamed, and when opened contained a suppurating appendix. With the extreme care warranted by the presence of sepsis, the appendectomy was successfully performed. Intense intra- and postoperative antibiotic therapy protected the patient from possible septic complications. This was the only instance that I encountered such an undeniably rare finding.

Rare pathologies of the appendix. Mention was made above of the involvement of the appendix by Crohn's Disease. This, however, is more commonly involvement of the appendix in the typhlitis caused by Crohn's (Fig. 4). On the contrary, the isolated localization of the disease in the organ is quite rare. The risks that appendectomy might entail in these situations were mentioned earlier. These are compounded all the more in the event the secondary nature of the appendicular disease is not distinguished from the cecal condition: the risk of fistulization.

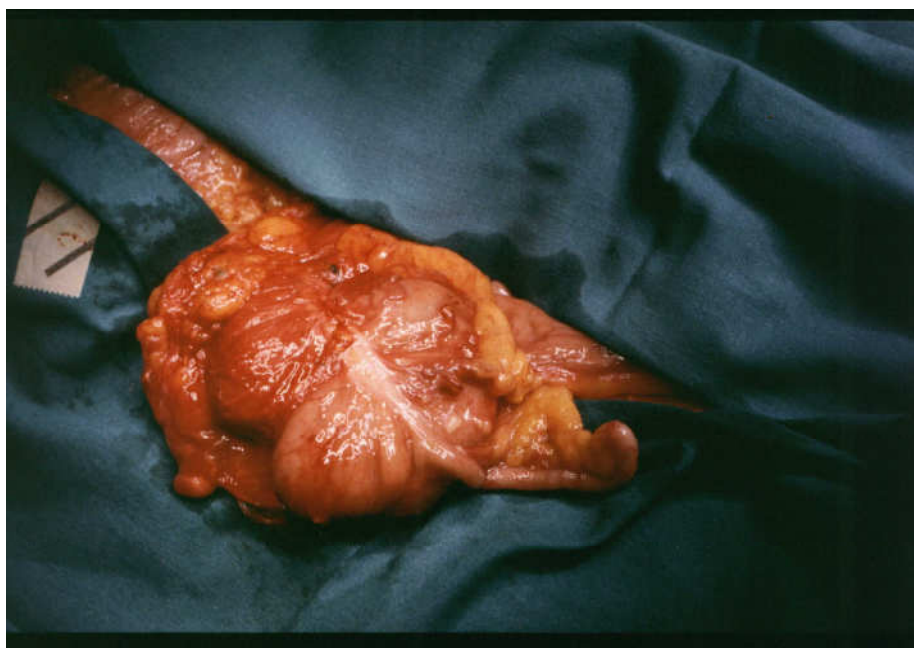


Fig. 4
Appendix in cecum affected by Crohn's Disease

The following are rare diseases of the appendix:

- Diverticulosis and diverticulitis of the appendix;
- Tubercular appendicitis;
- Appendicular mucocele;
- Appendicular oxyuriasis;
- Appendicular invagination
- Ileo-cecal-colic membrane

Tumors, too, are among rare diseases of the appendix. Exceptional neoplasms, both benign - adenomas, schwannomas, etc. - and malignant - adenocarcinomas (Fig.s 5 , 6), sarcomas, etc., have been described.

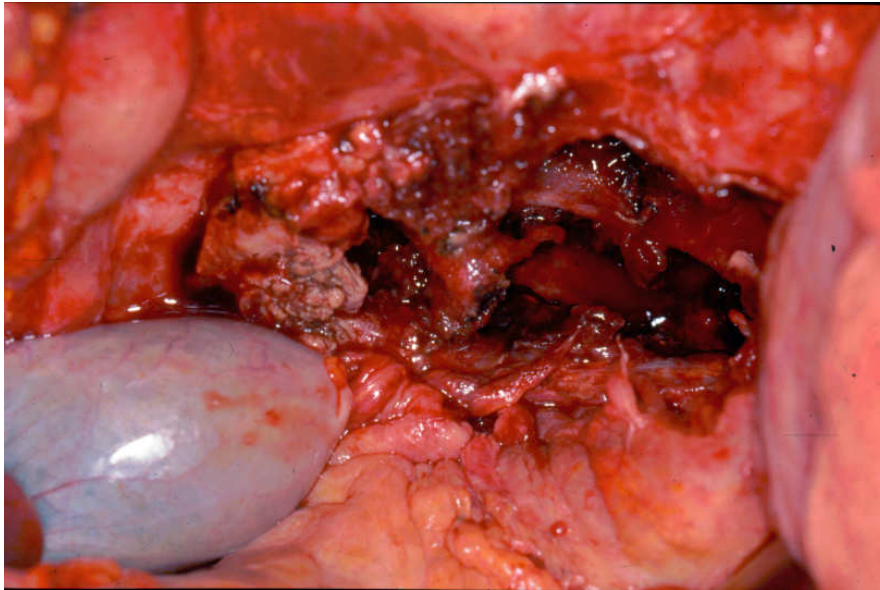


Fig. 5
Vast neoplastic source originating from the appendix

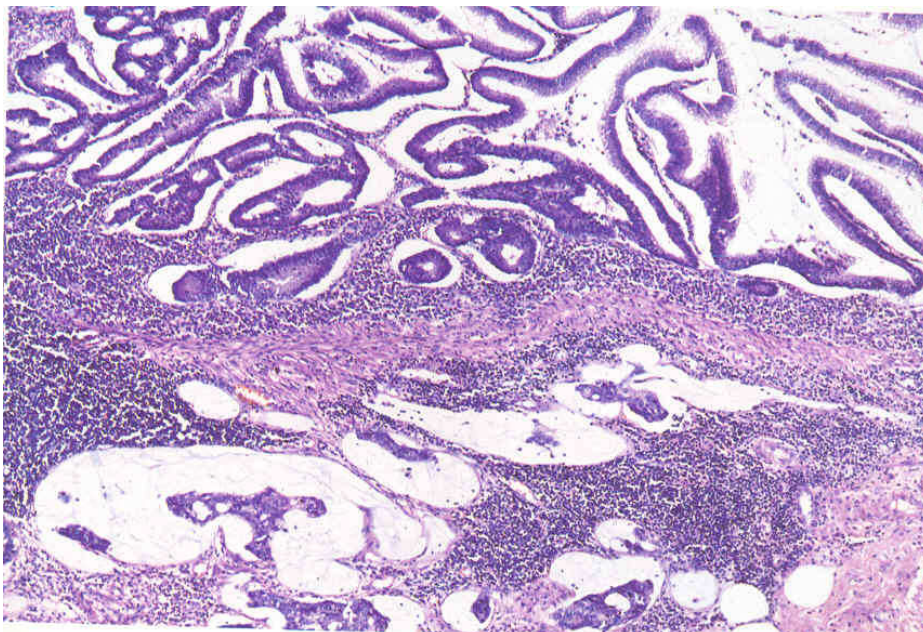


Fig. 6
Mucosal adenocarcinoma

It is worthwhile to briefly touch on the neuroendocrine tumor known as carcinoid (Fig.7), which is found fairly often in the appendix, by far more so than other tumors.

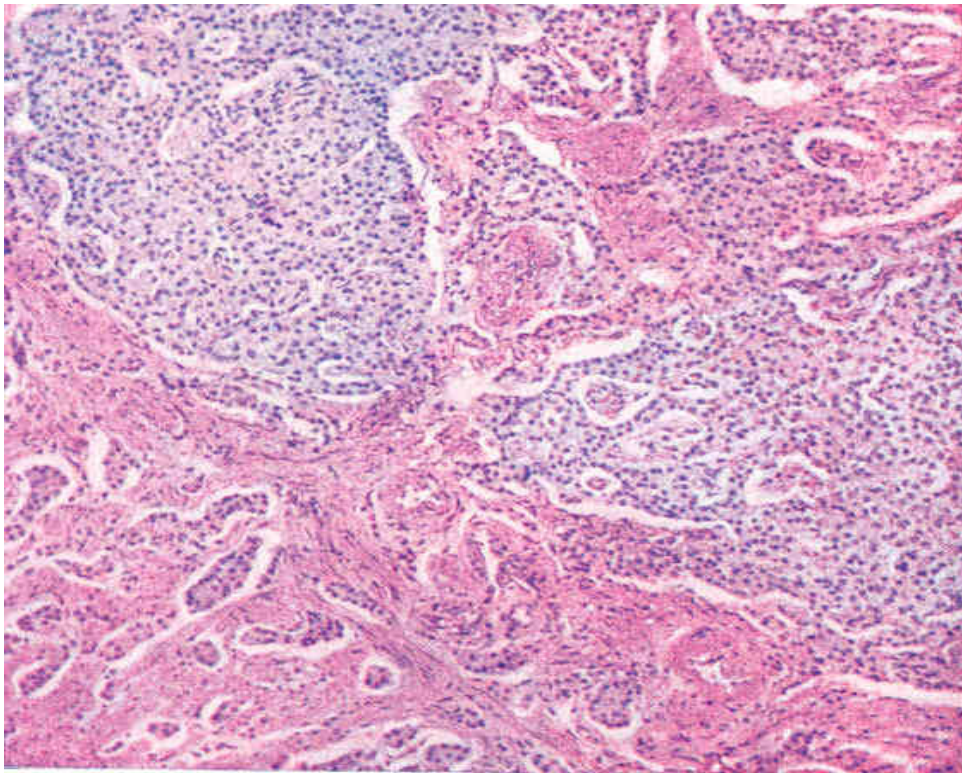


Fig. 7
Carcinoid tumor of the appendix

This often goes unnoticed, especially if the excised appendix is not subject to histopathologic examination. It must be said that the lesion does not usually display the vasoactive properties (“*carcinoid syndrome*”) commonly associated with this tumor, nor does it assume a malignant evolution (except in very rare cases). A carcinoid tumor is normally limited in size (when it is not only a histological specimen), and the symptomatology can generate pain, but this is dependent on the obstructive mechanisms that it creates. Indeed, intervention is generally undertaken for the appendicular disturbances induced by obstruction of the appendicular lumen that this lesion, however small it may be, can cause.

In conclusion, we can say that, despite the organ’s apparent exiguity, appendicular disease can represent a dangerous condition for the patient and the source of diagnostic difficulties - and at times also of errors - for the physician. Suffice it to recall position anomalies of the intestine (Lecture 23), the risk that appendicular inflammation may become systemic, the insidiousness of often overlooked chronic appendicitis and, finally, the albeit rare presence of neoplastic structures.

It is hoped that new and future generations of surgeons will be able to draw helpful insight from the experience gained over years of practice with this often cunning disease.

Videos

1. Laparoscopic appendectomy - Technique: Loop
2. Laparoscopic appendectomy - Technique: Clips - Endo GIA stapler
3. Acute abdomen - Acute appendicitis
4. Acute abdomen - Phlegmonous appendicitis with severe peritonitis